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TESIS DOCTORAL

TRANSICIÓN EN NIÑOS Y ADOLESCENTES CON ENFERMEDADES REUMÁTICAS

Daniel Clemente Garulo

Directores

Francisco López Muñoz

Loreto Carmona Ortells

Tutor

Francisco López Muñoz

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Tesis doctoral presentada por el licenciado en Medicina y Cirugía **D. Daniel Clemente Garulo**, para optar al grado de Doctor por la Universidad Camilo José Cela (Programa de Doctorado en Ciencias de la Salud).

DIRECTORES:

Francisco López Muñoz

Director de la Escuela Internacional de Doctorado de la Universidad Camilo José Cela

Loreto Carmona Ortells

Directora científica del Instituto de Salud Musculoesquelética

TUTOR:

Francisco López Muñoz

Director de la Escuela Internacional de Doctorado de la Universidad Camilo José Cela

Universidad Camilo José Cela

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Que, D. Daniel Clemente Garulo, licenciado en Medicina y Cirugía por la Universidad de Alcalá, ha realizado, bajo nuestra dirección, el trabajo de investigación titulado "TRANSICIÓN EN NIÑOS Y ADOLESCENTES CON ENFERMEDADES REUMÁTICAS". Consideramos se trata de un trabajo bien elaborado, de actualidad, que reúne todas las condiciones necesarias y de calidad suficiente, que autorizamos a ser defendido en lectura pública como TESIS DOCTORAL.

En Madrid, a 9 de Abril de 2018.

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Dra. Dª. Loreto Carmona Ortells

"It is easier to build strong children than to repair broken men"

"Es más fácil construir niños fuertes que reparar adultos rotos"

Frederick Douglass

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ÍNDICE

LISTADO DE ABREVIATURAS Y SIGLAS

AIJ artritis idiopática juvenil.

AINE antiinflamatorio no esteroideo.

ANA anticuerpos antinucleares.

ANCA anticuerpos anticitoplasma de neutrófilo.

ENA antígenos nucleares extraíbles (del inglés *extractable nuclear antigens*).

ERyME Enfermedades reumáticas y musculoesqueléticas.

EULAR *European League Against Rheumatism*.

FR factor reumatoide.

HLA antígeno leucocitario humano (acrónimo del inglés *Human Leukocyte Antigen*).

ILAR *International League of Associations for Rheumatology*.

LES lupus eritematoso sistémico.

OMS Organización Mundial de la Salud.

PReS *Pediatric Rheumatology European Society*.

RM resonancia magnética.

TC tomografía computarizada.

LISTADO DE TABLAS Y FIGURAS

RESUMEN

El objetivo general de esta investigación es poner de manifiesto los problemas durante el proceso de transición de adolescentes y jóvenes con enfermedades reumáticas y sugerir indicaciones para llevarla a cabo. Proporcionar una atención dirigida durante la adolescencia y asegurar la continuidad de su asistencia en las unidades de reumatología es fundamental para que sean capaces de conseguir un funcionamiento óptimo en la vida adulta.

Para ello, se realizó una evaluación de la situación actual de la transición en adolescentes y jóvenes con enfermedades reumáticas mediante una encuesta a reumatólogos pediátricos europeos con preguntas dirigidas acerca de su práctica real y una descripción de los programas de transición en reumatología mediante una revisión sistemática. De este modo, se identificaron las estrategias y herramientas más relevantes para el desarrollo de los programas y se elaboraron una serie de recomendaciones mediante un proceso de consenso para mejorar la asistencia durante el proceso de transición.

Los programas de transición de mayor calidad contienen seis puntos críticos: un programa de transición estructurado y por escrito; una planificación individualizada del proceso, con flexibilidad en su implementación y duración; un comienzo precoz; un coordinador que asuma la responsabilidad de la transición; la promoción de conocimientos y habilidades a jóvenes y sus padres; y la transmisión de la información relevante y traslado a la unidad de adultos. Sin embargo, faltan indicadores estandarizados y medidas que permitan evaluar adecuadamente el proceso de la transición en jóvenes con enfermedades reumáticas.

Los resultados de nuestra encuesta demuestran limitaciones en la implantación real de la transición y una escasa disponibilidad de recursos. Aunque se considera necesario proporcionar una atención específica en la transición a la atención en las unidades de adultos, no todos los centros desarrollan un programa de transición y es más frecuente un proceso de transición informal que la aplicación de programas

estructurados. La variabilidad encontrada depende de factores locales como los sistemas de atención sanitaria, marcos regulatorios y la disponibilidad de financiación.

Por último, se elaboran 12 recomendaciones que proporcionan información relevante sobre las estrategias necesarias para alcanzar resultados óptimos en la transición de jóvenes con enfermedades reumáticas, según la evidencia disponible y la opinión de un grupo multidisciplinar de expertos. Dado el gran interés de los reumatólogos pediátricos y de adultos por mejorar el proceso de transición, se espera que estas recomendaciones sean útiles para cambiar la práctica actual.

Palabras clave: enfermedades reumáticas, adolescentes, transición a la vida adulta.

ABSTRACT

The general objective of this research is to highlight the problems during the transition process of adolescents and young people with rheumatic diseases to adulthood and suggest indications to carry it out. It is essential to provide an appropriate care during adolescence and assure continuity of care in rheumatology units in order to get an optimal performance in life.

To this end, an assessment of the current situation of the transition in adolescents and young people with rheumatic diseases was made through a survey of European pediatric rheumatologists with questions about their actual practice and a description of the transition programs in rheumatology through a systematic review. In this way, the most relevant strategies and tools for the development of the programs were identified and recommendations were elaborated through a consensus process to improve assistance during transition to adult care.

The highest quality transition programs contain six critical points: a structured and written transition policy; an individualized planning of the process, with flexibility in its implementation and duration; an early start; a coordinator who assumes responsibility for the transition; the promotion of knowledge and skills to young people and their parents; and the transmission of the relevant information and transfer to the adult unit. However, there is a lack of standardized indicators and measures to adequately evaluate the transition process in young people with rheumatic diseases.

The results of our survey demonstrate limitations of existing transition practices and paucity of resources. Although it is considered necessary to provide specific attention in the transition to adult care, not all centers develop a transition program and an informal transition process is more frequent than the application of structured programs. The variability found depends on local factors such as health care systems, regulatory frameworks and funding availability.

Finally, 12 recommendations that provide relevant information on the strategies necessary to achieve optimal results in the transition of young people with rheumatic

diseases according to the available evidence and the opinion of a multidisciplinary group of experts are made. Given the great interest by pediatric and adult rheumatologists for improving the transition process, these recommendations are expected to be useful in changing current practice.

Key words: rheumatic diseases, adolescents, transitional care

1 INTRODUCCIÓN

1.1 Enfermedades reumáticas en la infancia

Las enfermedades reumáticas constituyen un grupo muy diverso y complejo en el que se incluyen todas las enfermedades con afectación del tejido conectivo, principal componente del sistema musculoesquelético (articulaciones, músculos, tendones, hueso), y que también forma parte de otros órganos y sistemas como piel, ojos o vasos sanguíneos. Aunque es más frecuente su aparición en la edad adulta, algunas enfermedades reumáticas tienen su inicio durante la infancia o la adolescencia. Entre ellas, destacamos por su importancia a la artritis idiopática juvenil (AIJ) y al lupus eritematoso sistémico (LES).

1.1.1 Artritis idiopática juvenil

La artritis idiopática juvenil es la enfermedad reumática inflamatoria crónica más frecuente de la infancia y una causa importante de morbilidad a corto y largo plazo. No se trata de una única enfermedad, sino más bien de un término bajo el que se engloba a todas las artritis de causa desconocida que aparecen antes de los 16 años y que persisten durante más de 6 semanas (1). Dependiendo del número de articulaciones afectadas durante los primeros 6 meses de enfermedad, la presencia o no de manifestaciones extraarticulares y, en algunos casos, factores genéticos, se distinguen 7 categorías de AIJ según la *International League of Associations for Rheumatology* (ILAR) (Tabla 1) (2).

La distribución de la AIJ es mundial, encontrándose resultados muy variables respecto a incidencia y prevalencia de la enfermedad. En Europa se ha descrito una incidencia de 1,6-23/100.000 niños menores de 16 años y una prevalencia de 3,8-400/100.000 niños menores de 16 años. La edad de aparición y el sexo predominante es variable dependiendo de cada categoría de AIJ (Tabla 2) (1, 3).

Tabla 1. Clasificación de AIJ según ILAR (Edmonton 2001).

Categoría AIJ	Criterios Inclusión	Criterios Exclusión*
Oligoarticular (persistente, extendida)	Artritis en 1-4 articulaciones en los 6 primeros meses de enfermedad, con subcategorías: Persistente: nunca > 4 articulaciones Extendida: > 4 articulaciones afectadas en evolución posterior	a, b, c, d, e
Poliarticular FR+	Artritis en ≥ 5 articulaciones en los 6 primeros meses de enfermedad con 2 determinaciones de FR (IgM) + en un intervalo de 3 meses	a, b, c, e
Poliarticular FR -	Artritis en ≥ 5 articulaciones en los 6 primeros meses de enfermedad, con determinación de FR (IgM) – en un intervalo de 3 meses	a, b, c, d, e
Sistémica	Artritis en 1 o más articulaciones asociada a fiebre diaria durante al menos 2 semanas, objetivada 3 días, con uno o más de los siguientes: 1. Exantema eritematoso evanescente 2. Adenopatías 3. Hepatomegalia y/o esplenomegalia 4. Serositis	a, b, c, d
Artritis relacionada con entesitis	Artritis y entesitis o artritis o entesitis y al menos 2 de los siguientes: 1.Dolor a la palpación de articulaciones sacroilíacas o dolor inflamatorio lumbosacro 2. HLA B27+ 3. Comienzo en varón mayor de 6 años 4. Uveítis anterior aguda 5. Antecedentes de EA, ArE, sacroileitis asociada a EII, síndrome de Reiter o UAA en familiar de primer grado	a, d, e
Psoriásica	Artritis y psoriasis o artritis y al menos 2 de los siguientes: 1. Dactilitis 2. Pocillos ungueales u onicolisis 3. Psoriasis en familiar de primer grado	b, c, d, e
Indiferenciada	Artritis que no cumple los criterios de ninguna categoría o que cumple de dos o más categorías	No aplica

*Criterios de exclusión: a) psoriasis o antecedentes de psoriasis en el paciente o en un familiar de primer grado; b) Artritis en un paciente varón, HLA B27+, por encima de los 6 años; c) Espondiloartritis anquilosante (EA), artritis relacionada con entesitis (ArE), sacroileitis asociada a enfermedad inflamatoria intestinal (EII), síndrome de Reiter o uveítis anterior aguda (UAA) en el paciente o en un familiar de primer grado; d) Factor reumatoide (FR) en al menos 2 determinaciones separadas por un intervalo de 3 meses; e) Presencia de AIJ sistémica en el paciente.

Manifestaciones clínicas

La inflamación articular está presente en todas las categorías de AIJ, caracterizándose por la presencia de tumefacción o la combinación de dolor y limitación funcional, puede variar el tipo y la distribución de las articulaciones afectadas (Tabla 2).

Tabla 2. Características principales de las distintas categorías de AIJ.

CATEGORIA AIJ	% del total de pacientes con AIJ	Género	Edad de inicio	Patrón articular
Oligoarticular	30-60	♀ > ♂	Primera infancia	≤ 4 articulaciones Grandes articulaciones (rodilla, tobillo, muñeca)
Poliarticular FR+	2-7	♀ > ♂	Infancia tardía / adolescentes	≥ 5 articulaciones Simétrico, erosiones
Poliarticular FR –	10-25	♀ > ♂	2 picos: 2-4 años y 6-12 años	≥ 5 articulaciones Simétrico
Sistémica	5-15	♀ = ♂	Cualquier edad	Oligo o poliarticular
Artritis relacionada con entesitis	5-10	♂ >>♀	Infancia tardía/ adolescentes	Predominio miembros inferiores, sacroileitis
Psoriásica	2-10	♀ > ♂	2 picos: 2-4 años y 9-11 años	Asimétrico o simétrico de pequeñas articulaciones o grandes articulaciones
Indiferenciada	10-20	Variable	Variable	Variable

La dactilitis es la tumefacción en un dedo que sobrepasa los márgenes articulares y es característica de la artritis psoriásica. Se define entesitis como la inflamación en el punto de inserción de tendones, ligamentos o cápsulas articulares en el hueso y es característica de la artritis relacionada con entesitis, al igual que la afectación del esqueleto axial, especialmente de las articulaciones sacroiliacas.

La uveítis o inflamación de la capa media del ojo es la manifestación extraarticular más importante en los pacientes con AIJ y está presente en el 10-30% de los casos. Se necesitan unas revisiones periódicas por el oftalmólogo para descartar su

presencia y evitar la aparición de complicaciones (4). También pueden aparecer manifestaciones cutáneas como psoriasis o alteraciones ungueales (AIJ psoriásica) o exantemas (AIJ sistémica). Manifestaciones constitucionales como astenia, anorexia, pérdida de peso aparecen en la AIJ sistémica y en algunos casos de AIJ poliarticular.

Diagnóstico

No hay ninguna prueba de laboratorio o de imagen que confirme por si misma el diagnóstico de AIJ, siendo necesario excluir otras causas conocidas de artritis. La determinación de anticuerpos antinucleares (ANA) es útil para valorar el riesgo de uveítis en pacientes con AIJ, no siendo específicos ni diagnósticos de enfermedad reumática. La determinación del factor reumatoide (FR) y del HLA B27 tampoco sirve para el diagnóstico de la enfermedad, sino para clasificar a un paciente ya diagnosticado en una categoría determinada. El análisis del líquido articular muestra un patrón inflamatorio sólo distinguible de las artritis sépticas mediante su cultivo.

Dentro de las pruebas de imagen, la ecografía articular es especialmente útil en el manejo de los pacientes con AIJ, ya que es capaz de detectar la presencia de sinovitis, tenosinovitis, entesitis y lesiones quísticas articulares o periarticulares. También permite la realización guiada de procedimientos como artrocentesis e infiltraciones.

La radiografía simple permite descartar causas ortopédicas y tumorales, así como valorar secuelas en los casos más evolucionados (erosiones, disminución espacio articular). La imagen por resonancia magnética es la técnica ideal para valorar determinadas articulaciones (temporomandibulares, caderas, articulaciones sacroiliacas), permite el diagnóstico precoz de la AIJ y la detección de secuelas óseas en etapas más tempranas que con la radiografía simple (5).

Tratamiento

El tratamiento de la AIJ requiere un enfoque multidisciplinar en unidades especializadas e incluye un tratamiento farmacológico, terapia física y rehabilitación, e intervenciones psicosociales. Entre los principales fármacos utilizados encontramos antiinflamatorios no esteroideos (AINEs), corticoides, tanto por vía oral, intraarticular o

intravenosa, y los denominados “fármacos de acción lenta o modificadores de la enfermedad”, entre los que encontramos el metotrexato, la leflunomida y la sulfasalazina. En los casos refractarios se emplean tratamientos biológicos como los antagonistas del TNFα (etanercept, adalimumab), terapias anti IL-6 (tocilizumab), terapias anti IL-1 (anakinra, canakinumab) o inhibidores de la coestimulación linfocitaria (abatacept) (6, 7).

El tratamiento rehabilitador y fisioterapéutico debe adaptarse a la actividad inflamatoria de cada articulación y el nivel de desarrollo del niño. En los casos más evolucionados puede ser necesario cirugías ortopédicas correctoras o la realización de artroplastias.

Evolución y pronóstico

La AIJ sigue un curso crónico en el que se alternan periodos de actividad y de remisión clínica. El pronóstico a largo plazo es difícil de establecer en los primeros meses desde el diagnóstico, siendo necesario para ello al menos un seguimiento de 3-5 años. Algunos indicadores de mal pronóstico son la mayor gravedad o número de articulaciones activas al inicio de la enfermedad, un patrón de afectación articular simétrico, la afectación de muñeca, cadera y/o tobillo, la presencia de FR, una actividad inflamatoria persistente o cambios radiológicos en etapas tempranas (1, 3).

1.1.2 Lupus eritematoso sistémico juvenil

El lupus eritematoso sistémico (LES) (8) es una enfermedad autoinmune crónica de causa desconocida ocasionada por la presencia de autoanticuerpos e inmunocomplejos que afectan a múltiples órganos y sistemas. Dada su similitud con la enfermedad en adultos, para establecer el diagnóstico pueden utilizarse los mismos criterios de clasificación del *American College of Rheumatology* (ACR) para LES, siendo necesaria la presencia simultánea o progresiva de ≥ 4 de los 11 criterios (Tabla 3) (9).

Tabla 3. Criterios de clasificación del *American College of Rheumatology* para lupus eritematoso sistémico.

Criterio	Definición
1. Eritema malar	Eritema fijo, liso o elevado, en "alas de mariposa", con tendencia a respetar surco nasogeniano
2. Eritema discoide	Placas eritematosas elevadas con hiperqueratosis; puede existir cicatrización atrófica en lesiones antiguas
3. Fotosensibilidad	Exantema tras la exposición solar, recogido en la historia o documentado por un médico
4. Úlceras orales	Ulceraciones orales o nasofaríngeas, no dolorosas
5. Artritis	Artritis no erosiva de articulaciones periféricas
6. Serositis	Pleuritis o pericarditis
7. Trastornos renales	Proteinuria persistente >0.5 g/dL o Cilindros celulares
8. Trastornos neurológicos	Convulsiones o psicosis en ausencia de causa metabólica o medicamentosa
9. Trastornos hematológicos	Anemia hemolítica con reticulocitosis o Leucopenia < 4000/µL en 2 o más ocasiones o Linfopenia < 1500/µL en 2 o más ocasiones o Trombocitopenia < 100000/µL
10. Trastornos inmunológicos	Anticuerpos antiADN o Anticuerpos antiSm o Anticuerpos antifosfolípido (anticuerpos anticardiolipina, presencia de anticoagulante lúdico o VDRL falso +)
11. Anticuerpos antinucleares	Por inmunofluorescencia o técnica equivalente

La incidencia anual en niños y adolescentes es de 0,36-0,9 por cada 100.000 niños y la prevalencia, muy variable según etnicidades es de 3,3-24 por cada 100.000 niños. La edad de aparición habitual es entre los 12-16 años, siendo rara por debajo de los 10 años. Es más frecuente en niñas (80%) y en asiáticos, afroamericanos y latinos.

La causa del LES sigue siendo desconocida, siendo fundamental en su desarrollo una disfunción del sistema inmune que aparece en pacientes predispuestos genéticamente y sobre el que actúan factores ambientales (luz ultravioleta, estrógenos, infecciones o fármacos).

Manifestaciones clínicas

Las manifestaciones clínicas son muy variables, pudiendo inicialmente presentar síntomas inespecíficos (fatiga, anorexia, pérdida de peso, cefalea) y asociar posteriormente manifestaciones específicas relacionadas según los órganos afectados, así como datos de inflamación sistémica (adenopatías, hepatoesplenomegalia. Las manifestaciones más frecuentes en LES juvenil son los síntomas constitucionales, la artritis, el exantema malar, las alteraciones hematológicas y la nefritis lúpica.

Alteraciones en los estudios complementarios

Las pruebas de laboratorio sirven para apoyar el diagnóstico de LES y para monitorizar la actividad de la enfermedad. La característica principal del LES es la presencia de múltiples autoanticuerpos, siendo los anticuerpos antinucleares (ANA) positivos (títulos > 1 / 160) en la mayoría de los pacientes. Los anticuerpos anti-DNA y los anti-Sm son muy específicos de LES. Otros anticuerpos que pueden encontrarse son los anti-SS-A (Ro) y anti-SS-B (La), los anti-RNP y los anticuerpos antifosfolípido. Para monitorizar la actividad de la enfermedad se utilizan los niveles de anti-DNA y los valores de complemento (C3 y C4), siendo estos últimos bajos o indetectables durante los periodos de actividad.

Tratamiento

El tratamiento del LES juvenil se basa en el uso de fármacos y en la atención a medidas generales de salud. El tratamiento farmacológico debe individualizarse según la extensión y gravedad de la enfermedad, incluyendo AINEs, corticoides tópicos y/o generalmente sistémicos (habitualmente durante periodos prolongados), hidoxicloroquina, metotrexato e inmunosupresores (ciclofosfamida, azatioprina). En los casos refractarios, pueden utilizarse inmunoglobulinas intravenosas, plasmaféresis o terapias biológicas dirigidas contra las células B, como rituximab (anticuerpo monoclonal anti CD-20) o belimumab (anticuerpo monoclonal anti-BLyS).

Dentro de las medidas generales es muy importante limitar la exposición al sol (radiación ultravioleta), recomendando protección solar (SPF ≥ 30) a diario. Se debe recomendar una ingesta adecuada de calcio y suplementos de vitamina D, controlar la dieta cuando se inicia un tratamiento con corticoides a dosis altas para evitar una excesiva ganancia ponderal y controlar y tratar los factores de riesgo cardiovascular, especialmente el hábito tabáquico (10).

Evolución y pronóstico

El LES es una enfermedad crónica que cursa con periodos de actividad y remisión, pudiendo desencadenarse exacerbaciones por factores exógenos como la exposición solar, infecciones, intervenciones quirúrgicas, etc. Las formas juveniles tienen peor pronóstico que los casos de inicio en la edad adulta, ya que presentan manifestaciones clínicas más graves y la remisión a largo plazo es poco frecuente, siendo más propensos a presentar complicaciones. Son factores de mal pronóstico la afectación orgánica grave, el género masculino, el origen étnico afroamericano, asiático o latino y un nivel socioeconómico bajo.

1.1.3 Otras enfermedades reumáticas de inicio en la infancia

Dermatomiositis juvenil

La dermatomiositis juvenil (DMJ) (11) es una enfermedad autoinmune caracterizada por la inflamación crónica de piel y músculo estriado. La presentación habitual de la DMJ es un cuadro de debilidad muscular de predominio en cintura escapular y pelviana asociado a un cuadro constitucional (fiebre, hiporexia, pérdida de peso) de comienzo insidioso y manifestaciones cutáneas específicas como las pápulas de Gottron o el eritema heliotropo. En casos graves puede aparecer disfagia o disnea por afectación de la musculatura faríngea y respiratoria. Durante su evolución pueden aparecer calcinosis, lipodistrofia y úlceras cutáneas. El tratamiento se basa en la administración de corticoides a dosis altas combinado con metotrexato, asociando inmunoglobulinas intravenosas en los pacientes con afectación moderada-grave. Como

terapia adyuvante de las lesiones cutáneas puede utilizarse tratamientos tópicos (corticoides, tacrólimos, pimecrólimus) o asociar hidroxicloroquina oral. A largo plazo, la mayor parte de los pacientes realiza una vida normal, aunque no todos puedan realizar actividad física intensa y hasta un 5% puede necesitar una silla de ruedas para desplazarse. En un 25-30% quedan contracturas articulares, atrofia muscular, como secuelas. Con los tratamientos actuales la mortalidad se ha reducido considerablemente (solo un 1-2 % de los casos, normalmente debido a complicaciones respiratorias.

Esclerodermia pediátrica

La esclerodermia (12) agrupa a un conjunto de enfermedades caracterizadas por la fibrosis o excesivo depósito de colágeno en la piel y en otros tejidos, diferenciándose formas localizadas o sistémicas, dependiendo de la localización y extensión de la fibrosis.

La esclerodermia localizada es la forma de esclerodermia más frecuente en la infancia, caracterizándose por la afectación de la piel y tejidos subyacentes, distinguiéndose diversos subtipos según la profundidad y el patrón de afectación de las lesiones. En las lesiones circunscritas superficiales con signos de actividad puede realizarse tratamiento tópico con corticoides, inhibidores de la calcineurina o fototerapia con luz ultravioleta. En los casos graves o refractarios se utilizan corticoides orales asociados a metotrexato o micofenolato mofetilo. El pronóstico va a depender de la superficie cutánea afectada, su localización y del grado de induración, así como de las secuelas articulares (contracturas, dismetría de extremidades, limitación de la función) y neurológicas y el impacto psicológico de la enfermedad.

La esclerosis sistémica es una enfermedad muy rara en niños, caracterizada por la afectación de piel, vasos y órganos internos. El inicio de la enfermedad se caracteriza por el desarrollo de un fenómeno de Raynaud asociado a edema e induración progresiva en manos y cara, telangiectasias (cara y extremidades superiores) y alteraciones capilares en el lecho ungueal. Las principales manifestaciones extracutáneas son gastrointestinales (reflujo gastroesofágico, trastornos en la motilidad digestiva), respiratorias (alteraciones en las pruebas de función pulmonar, enfermedad intersticial pulmonar, hipertensión pulmonar) y musculoesqueléticas (artralgias y artritis, sobre

todo en manos). El metotrexato y el micofenolato han demostrado mejorar la clínica cutánea, mientras que para la enfermedad pulmonar intersticial se precisa el uso de ciclofosfamida. Para el resto de las manifestaciones clínicas suele realizarse un tratamiento sintomático. Aunque la afectación cutánea y las contracturas articulares pueden dar a una discapacidad funcional grave, el pronóstico está determinado por la afectación cardiopulmonar y renal.

Enfermedad mixta del tejido conectivo

La enfermedad mixta del tejido conectivo (EMTC) se caracteriza por presentar signos y síntomas de dos o más enfermedades reumáticas y la presencia de anticuerpos anti-RNP. Las manifestaciones clínicas iniciales más frecuentes son la presencia de un síndrome constitucional asociado a fenómeno de Raynaud y la poliartritis de manos, apareciendo progresivamente manifestaciones de AIJ (sinovitis), LES (exantema malar, pleuritis, pericarditis, adenopatías), DMJ (debilidad muscular, miositis) y esclerodermia pediátrica (esclerodactilia, enfermedad pulmonar intersticial, trastornos en la motilidad esofágica). La mayoría de los pacientes suelen responder a corticoides a dosis bajas, antiinflamatorios no esteroideos (AINEs), hidroxicloroquina o combinaciones de estas medicaciones. El pronóstico a largo plazo es variable, siendo peor para los casos que se asemejan a LES y especialmente si asocian trombocitopenia e insuficiencia renal.

Síndrome de Sjögren

El síndrome de Sjögren se caracteriza por la inflamación de las glándulas exocrinas, principalmente glándulas salivales y lagrimales. En las formas juveniles la forma de presentación habitual es la parotiditis recurrente (75% casos), apareciendo posteriormente sequedad oral (dificultad para salivar durante las comidas o para hablar) y ocular además de otras manifestaciones sistémicas variables. La sospecha diagnóstica se refuerza con la presencia de ANA y anticuerpos anti-SSA o anti-SSB en la analítica y puede confirmarse mediante el estudio anatomopatológico de biopsias de glándulas salivares menores, que demuestra un infiltrado inflamatorio linfocitario. El tratamiento suele ser sintomático: lágrimas artificiales, estimulantes de la salivación, una buena

higiene dental y antiinflamatorios no esteroideos para los dolores articulares. Para el tratamiento de las manifestaciones sistémicas se utiliza hidroxicloroquina o metotrexato. Es importante tener en cuenta durante su seguimiento que existe un riesgo aumentado de linfoma en estos pacientes.

Vasculitis

Las vasculitis son un grupo heterogéneo de enfermedades multisistémicas caracterizadas por la presencia de inflamación en la pared de los vasos sanguíneos. En 2005, la *European League Against Rheumatism* (EULAR) y la *Pediatric Rheumathology European Society* desarrollaron la primera clasificación de vasculitis pediátricas según el calibre de los vasos afectados y la presencia o no de granulomas en el estudio histológico (Tabla 4) (13).

Tabla 4. Clasificación EULAR/PReS para vasculitis pediátricas.

Predominio de vasos grandes	Púrpura de Schönlein Henoch
Arteritis de Takayasu	Vasculitis leucocitoclástica cutánea aislada
Predominio de vasos medianos	Vasculitis urticaria hipocomplementémica
Poliarteritis nodosa	
Poliarteritis cutánea	
Enfermedad de Kawasaki	**Otras vasculitis**
	Enfermedad de Behcet
Predominio de vasos pequeños	Vasculitis asociada a infecciones, neoplasias o medicamentos
Granulomatosas	Vasculitis asociada a enfermedades del tejido conectivo
Granulomatosis de Wegener (granulomatosis con poliangeitis)	Síndrome de Cogan
Síndrome de Churg-Strauss	No clasificables
No granulomatosas	
Poliangeítis microscópica	

Son enfermedades raras, siendo la púrpura de Schönlein Henoch y la enfermedad de Kawasaki las vasculitis pediátricas más frecuentes.

Las manifestaciones clínicas y su pronóstico van a depender de la localización, la extensión y el calibre de los vasos afectados y, en las formas secundarias, de la causa subyacente. Debe sospecharse una vasculitis ante una fiebre de origen desconocido que asocia alteraciones cutáneas (púrpura, nódulos dolorosos, úlceras, lívedo reticular), neurológicas (neuropatía periférica), musculoesqueléticas (artralgias, artritis, miositis), renales o pulmonares.

Cuando la sospecha clínica es alta, la realización de una angioRM, un angioTC o una angiografía convencional puede demostrar alteraciones vasculares (estenosis, aneurismas), sobre todo en las vasculitis que afectan a vasos de mediano o gran calibre. El diagnóstico de confirmación se realiza mediante el estudio anatomopatológico, tras la biopsia de los tejidos afectados.

El tratamiento y la evolución varían dependiendo del tipo de vasculitis, incluyendo AINEs, corticoides, inmunoglobulinas intravenosas e inmunosupresores (ciclofosfamida, micofenolato) (14).

Síndromes autoinflamatorios

Los síndromes autoinflamatorios agrupan a un conjunto de entidades que se caracterizan por episodios recurrentes de inflamación sistémica causadas por alteraciones en la regulación de la inmunidad innata, no encontrándose anticuerpos ni células T autorreactivas. En muchos casos se deben a mutaciones genéticas que dan lugar a un aumento de la síntesis de diversas citoquinas proinflamatorias (especialmente la IL-1β) responsables de las manifestaciones clínicas y analíticas.

Deben sospecharse estos síndromes en pacientes con fiebre recurrente no originada por otros cuadros y que asocie manifestaciones en varios sistemas, especialmente en piel, tracto digestivo, ojos, aparato locomotor y sistema nervioso central o en aquellos pacientes con elevación de los parámetros inflamatorios de forma inexplicada aun estando asintomáticos. En algunos casos pueden encontrarse antecedentes familiares. Dada la baja frecuencia de estos cuadros, el estudio genético sólo se realiza en los pacientes con alta sospecha clínica.

Los corticoides van a atenuar los síntomas en muchos casos, aunque de una forma temporal y limitada por los efectos adversos y suelen ser necesario asociar otros tratamientos, como la colchicina o los tratamientos biológicos dirigidos contra la IL-1. En cualquier caso, el tratamiento debe individualizarse y ser guiado por unidades especializadas dada la complejidad de los pacientes. El pronóstico va a depender de la aparición de amiloidosis renal, secundaria a la inflamación mantenida, así como de la extensión de los órganos y sistemas afectados (15).

1.2 La transición en niños y adolescentes con enfermedades reumáticas

1.2.1 Adolescencia y juventud

La adolescencia puede definirse como el periodo de la vida comprendido entre el final de la infancia y el inicio de la edad adulta. Según la Organización Mundial de la Salud (OMS) se considera adolescencia el periodo entre los 10 y los 19 años, pudiendo diferenciarse una adolescencia temprana (10-13 años), media (14-16 años) y tardía (17-19 años). Estos límites se entienden de forma flexible, pues hay evidencia de comienzo puberal antes de los 10 años y problemas que no se resuelven hasta mediada la tercera de cada de la vida. La juventud es un término más amplio que abarca el periodo entre los 10 y los 24 años, incluyendo adolescentes y adultos jóvenes (20-24 años) (16).

Esta etapa se caracteriza por ser una etapa de rápido desarrollo y de importantes cambios a nivel fisiológico, psicológico y social. No solo tiene lugar la maduración física y sexual, sino que durante estos años se desarrollan además la identidad personal, las vocaciones y las capacidades para interactuar socialmente y establecer relaciones de pareja, al mismo tiempo que se adquieren responsabilidades de forma progresiva y, generalmente, se busca la independencia respecto a los progenitores (17).

El patrón de desarrollo neurocognitivo durante la adolescencia determina que sea un periodo de mayor vulnerabilidad e inestabilidad emocional. Mientras que los centros responsables de las conductas impulsivas y los sistemas de recompensa están

desarrollados durante la adolescencia temprana, los cambios en la corteza prefrontal del cerebro, responsable del pensamiento abstracto y de la planificación a largo plazo, no finalizan hasta el final de la segunda década de la vida o incluso más adelante (18). De este modo, los adolescentes y jóvenes adultos son más vulnerables a la presión del grupo y al desarrollo de conductas de riesgo como trastornos de la conducta alimentaria (obesidad, anorexia nerviosa, bulimia), hábitos tóxicos (tabaco, alcohol, drogas), accidentes (disputas, agresiones, accidentes de tráfico), enfermedades de transmisión sexual o embarazos no deseados, etc. (19). Estas conductas de riesgo son más frecuentes todavía en aquellos adolescentes y jóvenes con enfermedades crónicas, reforzando la necesidad de actividades preventivas y de promoción de la salud en este grupo de edad (20).

La adolescencia y la juventud es un periodo de la vida complejo y de una duración cada vez mayor debido a cambios sociales durante los últimos años (p.ej., abandono más tardío del domicilio familiar) (21). Aunque la mayoría de los individuos superan este periodo sin experimentar grandes dificultades o dificultades permanentes, la relación con los adolescentes y jóvenes pueda ser complicada para los padres y los profesionales sanitarios que los atienden.

1.2.2 Impacto de las enfermedades reumáticas en la adolescencia

Las enfermedades reumáticas que aparecen en la infancia y la adolescencia tienen una gran influencia en todos los aspectos de la vida, tanto por la actividad de la enfermedad como por los tratamientos empleados, dando lugar a alteraciones físicas, psicológicas y sociales (Tabla 5). Dada la heterogeneidad de las enfermedades reumáticas el impacto sobre el desarrollo es variable, incluso mínimo en algunos casos como la AIJ de curso oligoarticular.

La talla baja es una complicación en algunos pacientes con AIJ, especialmente las formas poliarticulares y sistémicas. La etiopatogenia es multifactorial, influyendo la gravedad y duración de la actividad inflamatoria, factores nutricionales y el uso prolongado de dosis altas de corticoides (22). También pueden encontrarse alteraciones

del crecimiento localizado en las articulaciones con inflamación persistente, dando lugar a discrepancias en la longitud de extremidades, deformidades en valgo o micrognatia/asimetrías faciales, en el caso de que se afecten a las articulaciones temporomandibulares (23). Además de provocar un aumento de la fragilidad ósea y del riesgo de fracturas, cuando se reciben altas dosis de corticoides aparecen cambios corporales como aumento de peso y redistribución de los depósitos grasos, desarrollo de estrías cutáneas o hirsutismo. La limitación de la movilidad cuando la enfermedad está activa provoca debilidad y atrofia muscular, originando una pérdida de la condición física con una menor tolerancia al ejercicio y alteraciones de la movilidad y equilibrio (24, 25).

Al igual que en otras enfermedades crónicas, existe un riesgo de sobreprotección por parte de los padres o cuidadores que entorpece la adquisición de la independencia y autonomía propia de la adolescencia tardía (26). La incapacidad de los niños con enfermedades reumáticas para participar plenamente en todas las actividades deportivas o de ocio puede disminuir su autoestima, conducir al aislamiento social e impedir el establecimiento de relaciones amistad o pareja con otros adolescentes (27). Aunque los resultados académicos son similares a otros jóvenes, la tasa de desempleo es mayor en jóvenes con enfermedades reumáticas debido principalmente a una disminución de la capacidad física. Además, entre los que trabajan, existe discriminación por la enfermedad en un 25% de los casos (28).

Los trastornos del estado de ánimo, como la depresión, son más frecuentes en pacientes con artritis. De hecho, el 38% de los pacientes con AIJ y depresión presentan su primer episodio entre los 15 y los 25 años (29). En estos adolescentes se genera un sentimiento de impotencia ya que los tratamientos no siempre previenen la aparición de recaídas de la enfermedad, que suelen además determinar una mayor dependencia de los padres. Esta baja autoestima está relacionada con la aparición de dolor crónico en la edad adulta (29). Por último, los adolescentes con enfermedades reumáticas pueden no recibir información adecuada acerca de temas importantes como sexualidad (30), a pesar de que hasta un tercio de los pacientes con AIJ mantienen relaciones sexuales antes de llegar a la unidad de adultos (31). Sin embargo, aunque los pacientes

con enfermedades crónicas sean sexualmente activos como otros compañeros de su edad, tienen un mayor riesgo de problemas como enfermedades de transmisión sexual, embarazos no deseados o abusos sexuales (32).

Tabla 5. Impacto de las enfermedades reumáticas en adolescencia.

Alteraciones físicas e imagen corporal	
Talla baja Pubertad retrasada Alteraciones locales del crecimiento (p.ej., micrognatia, dismetría de extremidades)	Atrofia y debilidad muscular Cambios en la apariencia física (redistribución de depósitos grasos, estrías cutáneas, hirsutismo, etc.)
Alteraciones psicológicas y salud mental	
Baja autoestima Sensación de impotencia y pérdida de control	Dolor crónico Conductas de riesgo
Alteraciones sociales	
Dependencia física de los padres Incapacidad para participar en actividades deportivas y de ocio	Disminución de relaciones sociales Problemas laborales

1.2.3 Diferencias entre unidades pediátricas y de adultos

La atención en las unidades pediátricas concluye en torno a los 14-18 años, estableciéndose un límite de edad variable dependiendo del país, del centro sanitario y del tipo de atención recibida, pudiendo variar entre atención primaria y especializada. En España, la asistencia sanitaria en atención primaria finaliza a los 14 años, mientras que este límite se amplía hasta los 18 años para la atención especializada como parte del II Plan Estratégico de Infancia y Adolescencia 2013-2016 aprobado en abril 2013. En algunas enfermedades, como las enfermedades oncológicas, se puede prolongar el seguimiento en unidades pediátricas hasta los 21 años.

El cambio de unidades pediátricas a unidades de adulto puede suponer una dificultad añadida para los adolescentes y sus familias con enfermedades reumáticas. En los servicios pediátricos se establece una relación de varios años con el equipo sanitario que les atiende y el trato con los pacientes suele ser más familiar, con un ambiente más relajado. Normalmente existe una menor presión asistencial y se dispone de más tiempo para atender a todos los problemas de los niños, no solo los relacionados con la enfermedad y los tratamientos recibidos. También suele haber una mayor disponibilidad para la atención de consultas "sin cita" y una mayor atención a las técnicas de sedoanalgesia durante los procedimientos potencialmente dolorosos o generadores de ansiedad como las artrocentesis e infiltraciones.

Por el contrario, en las unidades de Reumatología no siempre son atendidos por un mismo profesional y la comunicación con el paciente es más formal y directa, pero puede percibirse como más distante. Dado el mayor número de pacientes atendidos el tiempo disponible en la consulta es menor y más centrado en los problemas relacionados con la enfermedad y los tratamientos, siendo más difícil la atención de los pacientes con problemas entre las citas programadas. También es menos frecuente la utilización de técnicas de sedoanalgesia (33, 34). Las principales diferencias entre unidades pediátricas y de adultos se recogen en la Tabla 6.

Además, el sistema sanitario de algunos países cubre sólo la financiación de determinados fármacos durante la etapa pediátrica, siendo posteriormente necesario la contratación de seguros médicos que aseguren la continuidad de los tratamientos (35, 36). Estas diferencias en la atención sanitaria pueden provocar que algunos pacientes afronten con inseguridad y ansiedad su traslado a las unidades de adulto, sobre todo cuando es necesario cambiar de centro. En algunos casos, esto puede suponer una pérdida en el seguimiento de los pacientes durante los primeros meses de su llegada a la unidad de Reumatología.

Tabla 6. Diferencias entre unidades pediátricas y de adultos.

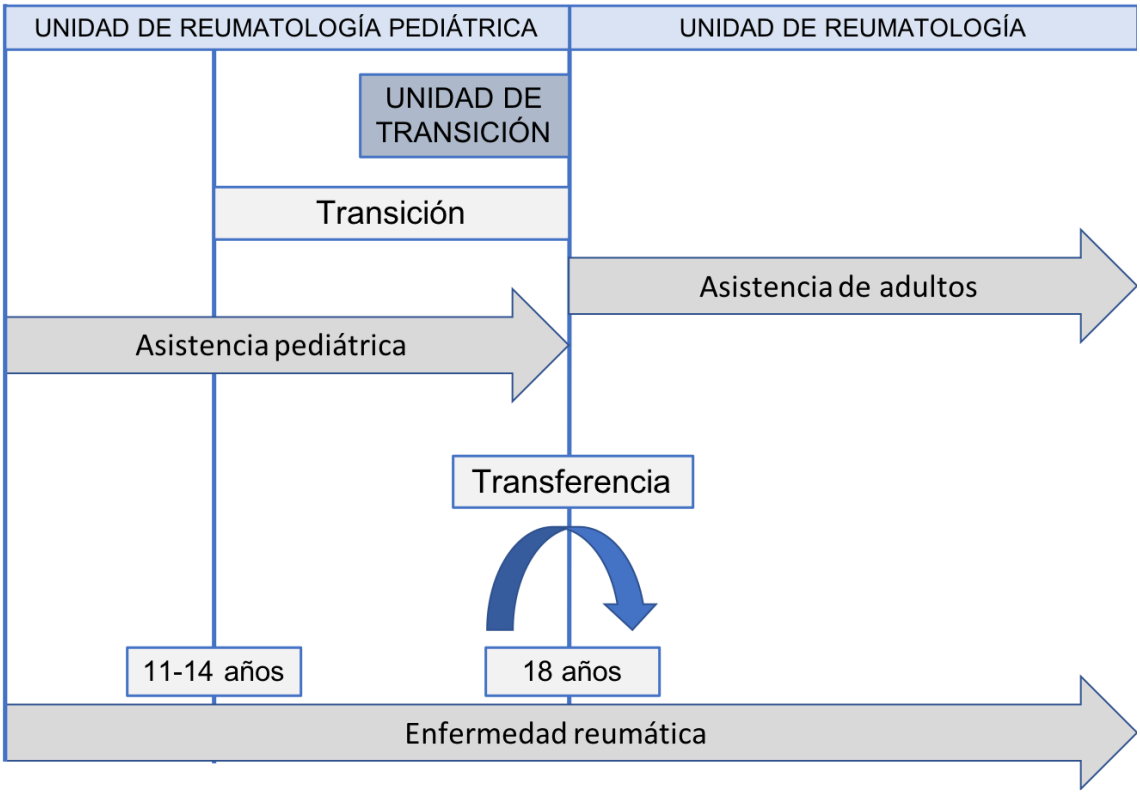
	Unidades pediátricas	Unidades en adultos
Consulta	Familiar, orientada a los padres	Individual, dirigida al paciente
Comunicación	Relajada e informal; empática pero también más paternalista	Estilo formal y directo, más distante, con mayor toma de decisiones por el paciente
Atención sanitaria	Holística; atención al desarrollo y al aprendizaje, así como al funcionamiento social Mayor supervisión	Centrada en la enfermedad, en las complicaciones del tratamiento y la adherencia al mismo
Equipo sanitario	Multidisciplinar, con mayor posibilidad de atención psicológica	Mayor dependencia de un especialista, con menor posibilidad de atención psicológica
Número de pacientes	Menor	Mayor
Tiempo de consulta	Mayor, permite obtener información más variada y detallada	Menor, permite obtener información menos información
Procedimientos de sedoanalgesia	Habituales	Menos frecuentes

1.2.4 El proceso de transición

La **transición** puede definirse como el proceso planificado de preparación, adaptación e integración paulatina de adolescentes y jóvenes con patologías crónicas en una unidad asistencial de adultos (37). La transición debe entenderse como un proceso activo y dinámico, capaz de abordar las necesidades médicas, psicosociales y educacionales/vocacionales de los adolescentes durante este periodo, siendo su objetivo final asegurar el desarrollo de habilidades y la adquisición de los recursos necesarios para afrontar con madurez e independencia su enfermedad. Este proceso se inicia habitualmente durante los primeros años de la adolescencia, debiendo adaptarse a la edad y el grado de desarrollo de los pacientes y finaliza cuando el paciente continúa su cuidado en las unidades de adultos. El evento administrativo en el que se traslada al paciente junto con la información clínica de un especialista pediátrico a otro adulto se

denomina **transferencia** y no debe confundirse con la transición (Figura 1) (38). Esta distinción no siempre está clara por parte algunos profesionales sanitarios, que ofrecen en sus centros programas de transición, pero que en realidad simplemente transfieren a los pacientes sin garantizar la adquisición de capacidades (39).

Figura 1. Atención durante la transición y transferencia.



La transición es especialmente relevante en los adolescentes y jóvenes con enfermedades reumáticas. A pesar de que durante las últimas décadas se han producido importantes avances en el tratamiento de estos pacientes, hasta un 40-50% de los casos los niños y adolescentes con enfermedades reumáticas presentan secuelas en la edad adulta o precisan continuar con tratamientos complejos, incluyendo el uso de inmunosupresores (40). Como se ha comentado anteriormente, también es habitual que el desarrollo psicosocial, sexual y vocacional se vea comprometido en comparación con sus compañeros (23, 31, 41). Por tanto, todos los adolescentes y jóvenes con enfermedades reumáticas deben recibir un cuidado continuo y una preparación adecuada para conseguir un funcionamiento óptimo en la vida adulta.

Sin embargo, a pesar del reconocimiento de la importancia de la transición, hay evidencia de que hasta la mitad de adolescentes y jóvenes con enfermedades reumáticas no logran acceder y continuar su seguimiento en las unidades de adultos de forma satisfactoria y presentan mayor riesgo de presentar complicaciones a largo plazo (42). Los factores que impiden alcanzar una transición exitosa pueden depender del paciente, de los padres/cuidadores o de los profesionales sanitarios (43-45).

En el caso de los pacientes, la necesidad de fármacos durante largos periodos de tiempo, las numerosas consultas médicas y el sentimiento continuo de sentirse diferente a los demás compañeros de su edad termina hastiando a estos jóvenes, que dan lugar a una falta de adherencia a los tratamientos pautados, absentismo en las revisiones programadas o la aparición de conductas de riesgo para su salud (p.ej., consumo excesivo de alcohol mientras recibe metotrexato, abandono del tratamiento con corticoides por el deterioro del aspecto físico). Estas situaciones son originadas por una falta de conocimientos sobre la enfermedad, de las consecuencias a largo plazo sin un tratamiento adecuado (muchas veces daños irreversible), y de la diferencia de prioridades entre el adolescente y la familia y/o los profesionales sanitarios que le atienden. Del mismo modo, puede resultar difícil convencer a un adolescente de la necesidad de un tratamiento cuando su enfermedad se encuentra inactiva, predisponiendo una nueva recaída y progresión de esta que puede llegar a ser difícil de controlar. Determinados comportamientos como la falta de puntualidad, el desinterés, o incluso actitudes desafiantes son circunstancias que afectan a la relación con los profesionales sanitarios.

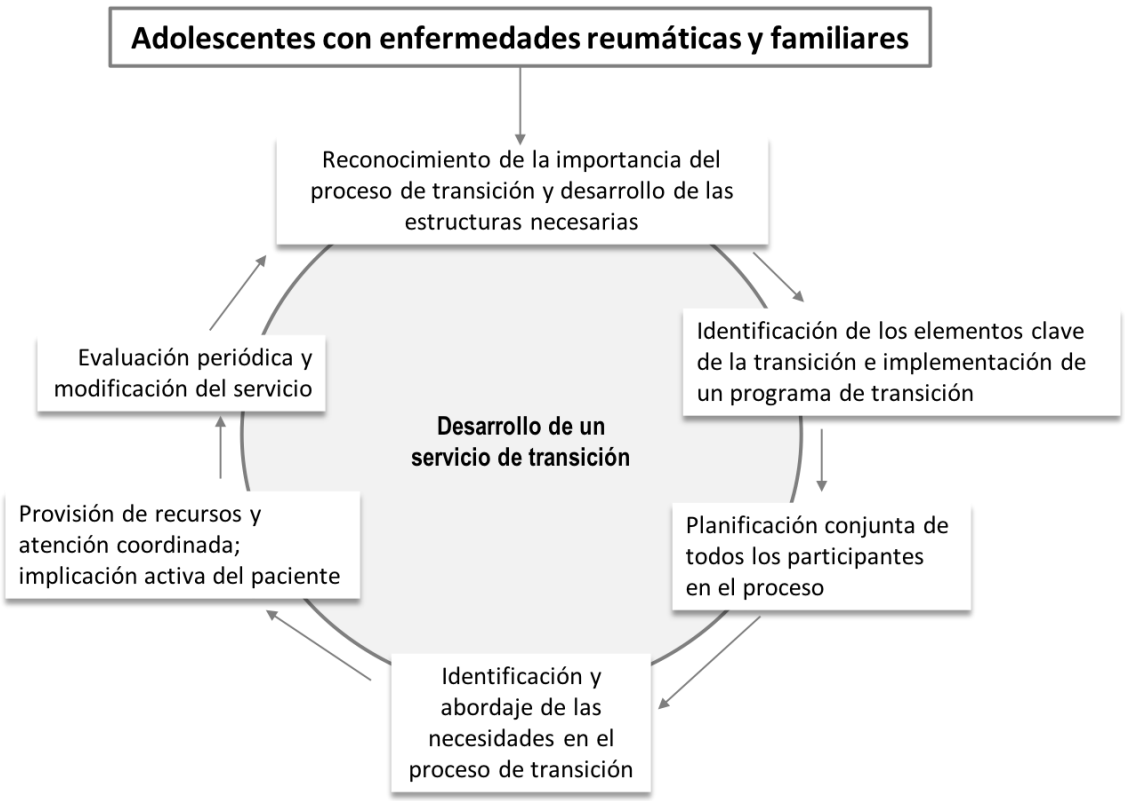
Por parte de los padres existe una reticencia a dejar que sus hijos participen en la toma de decisiones relativas a su tratamiento o las revisiones médicas. Otra limitación habitual en el proceso de transición, tanto por parte de los padres como de los jóvenes, es la dificultad para asumir el cambio de equipo y centro sanitario que conocen de muchos años.

Por último, muchos profesionales sanitarios no se sienten preparados para abordar con los adolescentes temas como sexualidad, consumo de alcohol y otras

drogas o problemas psiquiátricos (p.ej., ideación suicida). Esto puede deberse a un tiempo limitado en la consulta para abordar estos asuntos, pero principalmente a la falta de formación de los profesionales sanitarios en el cuidado del adolescente (46, 47). A esto se añaden los problemas de coordinación y comunicación entre unidades de reumatología pediátrica y de adultos y, en algunos países, de la falta de financiación sanitaria una vez terminado la etapa pediátrica.

Para superar estas dificultades y proporcionar una asistencia integral a los adolescentes y jóvenes con enfermedades reumáticas es necesaria la implicación activa del paciente, su entorno familiar/social y de un equipo multidisciplinar coordinado compuesto tanto de especialistas pediátricos como adultos. Para ello, varios centros han desarrollado e implementado distintos programas de transición (Figura 2).

Figura 2. Desarrollo de un programa de transición.



La reproductibilidad y el impacto real de estos programas en la práctica clínica es variable, dado que son dependientes de la organización del sistema sanitario

particular, la existencia o no de financiación para los mismos y el hecho de que no siempre se hayan utilizado medidas objetivas e indicadores de calidad apropiados para su evaluación.

Es por tanto necesario mejorar el proceso de transición para adolescentes y jóvenes con enfermedades reumáticas mediante el desarrollo de recomendaciones y guías de actuación específicas, así como de establecer indicadores de calidad y medidas objetivas para evaluar los programas de transición.

2 Objetivos e hipótesis

El **objetivo general** de la tesis es poner de manifiesto los problemas de la transición en adolescentes con enfermedades reumáticas y sugerir directrices.

Los objetivos específicos de la tesis son:

1) Evaluar la situación actual de la transición en adolescentes y jóvenes con ER.
2) Describir los programas de transición existentes.
3) Elaborar estándares y recomendaciones para mejorar la asistencia en las unidades de transición y asegurar un cuidado adecuado de estos pacientes, y establecer indicadores de calidad y medidas objetivas de resultados para promover la investigación en transición y la práctica basada en la evidencia.

Se establecieron **hipótesis** para los objetivos 1 y 2, pero no para el objetivo 3, dada la naturaleza del proyecto, de corte cualitativo.

Las hipótesis fueron:

1) Existen programas establecidos validados que abordan la transición basándose en evidencia y en opinión; algunos de estos programas, pero no todos, han sido evaluados.
2) Existe una gran variabilidad en el abordaje de la transición en Europa y no siempre se hace mediante un programa formal o estrategia con efectividad evaluada.

3 Metodología

Toda la metodología y el desarrollo de los distintos estudios que conforman la tesis se llevaron a cabo con la supervisión del grupo de trabajo para la Transición de la *European League Against Rheumatism* (EULAR) / *Pediatric Rheumatology European Society* (PRES).

La metodología de la tesis incluyó: revisión sistemática, encuesta a profesionales y metodología de consenso.

3.1 Revisión sistemática

Se llevó a cabo una revisión sistemática de programas de transición validados e implementados en la vida real, describiendo los elementos que contienen y prestando especial atención a potenciales recomendaciones, estándares, estrategias para su implantación y herramientas para la evaluación de resultados.

Una vez seleccionados, los artículos se analizaron utilizando métodos estándar para revisiones sistemáticas (http://www.prisma-statement.org/). La información obtenida se utilizó para respaldar una iniciativa de consenso para definir potenciales estándares y recomendaciones para mejorar la atención durante la transición.

3.1.1 Estrategia de búsqueda

Se usaron Medline, Embase y PsycINFO (todos desde el inicio hasta octubre de 2014) para las búsquedas, incluyendo términos libres y sinónimos de términos MeSH de "ERyME", "cuidado de transición" y "resultados". La búsqueda se limitó a adolescentes, niños y adultos jóvenes y a estudios publicados en inglés, alemán y español. Además, escaneamos las listas de referencias y la función "Artículos relacionados" en PubMed (www.pubmed.gov) de los artículos incluidos, y buscamos los resúmenes de las reuniones ACR, EULAR y PRES disponibles en sus sitios web oficiales.

3.1.2 Selección de estudios

Los estudios se seleccionaron si describían programas de transición "validados" en ERyME juveniles. El término "validado" se refería a programas que se habían implementado y probado en condiciones de la vida real de acuerdo con indicadores o resultados previamente especificados (es decir, el programa se había descrito con suficiente detalle como para ser reproducido). Dos revisores examinaron de forma independiente los resultados obtenidos por las búsquedas de artículos potencialmente relevantes; primero escaneando el título y el resumen, y luego mediante una revisión detallada del texto completo. Aquellos artículos que cumplían con los criterios de inclusión fueron evaluados críticamente e independientemente por los revisores, extrayendo la siguiente información sobre los programas de transición: país y entorno del programa, enfermedades y edades de los pacientes, recursos y personal disponible, figura de coordinador de la transición, lugar de consulta, proceso de transición, indicadores disponibles y resultados de efectividad. Cualquier discrepancia se resolvió por consenso con un metodólogo.

3.1.3 Evaluación de calidad

En ausencia de una herramienta apropiada disponible para revisiones sistemáticas de los servicios de salud, definimos una escala *ad hoc* para calificar la calidad de los programas según las siguientes dimensiones: el nivel de definición o reproducibilidad, si el programa se basó en investigaciones previas, la disponibilidad de indicadores de calidad y la presencia de resultados o evidencia de la efectividad del programa.

3.2 Encuesta

Se realizó un estudio exploratorio acerca de cómo se realiza la transición en adolescentes y jóvenes con enfermedades reumáticas en la práctica clínica real mediante una encuesta a reumatólogos pediátricos europeos.

Se invitó a los reumatólogos pediátricos europeos a completar una encuesta electrónica anónima de 17 ítems para evaluar su práctica de transición actual. El cuestionario fue desarrollado después de la revisión sistemática de la literatura y una evaluación crítica de los programas de transición en reumatología y dos reuniones presenciales del grupo de trabajo PReS / EULAR para la transición. Se incluyeron elementos que indagaban sobre las prácticas de transición actuales, los elementos clave de los programas de transición y los recursos disponibles. El cuestionario incluyó elementos sobre el servicio y el desarrollo del proceso de transición, el personal y los recursos disponibles.

El estudio se realizó en cooperación con PRINTO. PRINTO es una red internacional de centros académicos y clínicos que participan activamente en la investigación / atención clínica de niños y adolescentes con enfermedades reumáticas. Los encuestados participaron voluntariamente en el estudio dando su consentimiento para la recopilación de datos con el propósito de la investigación científica. La encuesta fue distribuida por el Centro Coordinador de PRINTO (Organización Internacional de Ensayos de Reumatología Pediátrica) a través de SurveyMonkey© a los directores de PRINTO de todos los centros en 25 países europeos. Se envió un recordatorio para aumentar la tasa de respuesta.

3.3 Metodología de consenso

El proceso de consenso se sometió a las siguientes etapas: 1) Establecimiento de un panel de expertos internacionales para incluir pacientes y representantes de equipos multidisciplinares de reumatología pediátrica y de adultos; 2) Una revisión sistemática de la literatura; 3) Establecimiento de un marco de trabajo, desarrollo de un mapa del proceso y generar un primer borrador de estándares y recomendaciones; 4) Iteración adicional de recomendaciones; 5) Desarrollo de recomendaciones de consenso con la metodología Delphi; y 6) Establecimiento de estándares e indicadores de calidad, como lo sugieren los Procedimientos Operativos Estándar de EULAR(48).

Los coordinadores del proyecto coordinaron el nombramiento de becarios clínicos para trabajar en el proyecto. Luego convocaron un panel multidisciplinario

experto de reumatología pediátrica y de adultos en toda Europa (médicos y profesionales de la salud relacionados con interés en la transición) y representantes de pacientes (adultos jóvenes con ERyME juveniles). El objetivo era que el panel reflejara la diversidad de Europa a nivel geográfico, cultural y de sistemas de atención médica.

Se utilizó la revisión sistemática de la literatura de los modelos existentes de atención transicional previamente explicada.

La primera reunión presencial del panel de expertos discutió los resultados de la revisión sistemática y acordó lo siguiente:

1) propósito del proyecto, cronogramas, roles y resultados
2) el 'mapa de proceso' de la transición utilizando el software MindManager®
3) un borrador con una propuesta de recomendaciones y estándares relacionados con diferentes elementos del proceso de transición; la lista propuesta se distribuyó después de la reunión por correo electrónico al grupo de expertos y se solicitaron más comentarios.

Una segunda reunión presencial definió mejor las recomendaciones y estándares en función de los comentarios del grupo. También se sugirieron indicadores de calidad apropiados para cada recomendación y estándar.

Las recomendaciones y estándares se enumeraron luego como afirmaciones. A continuación, se invitó a una audiencia más amplia de un total de 195 profesionales sanitarios a participar en una encuesta *on-line* y dar su opinión sobre el nivel de acuerdo con cada afirmación. La encuesta electrónica se difundió a través de listas de correo electrónico mantenidas por grupos profesionales (como PReS, EULAR y sociedades de reumatología en diferentes países). Todas las respuestas fueron anonimizadas. A los participantes se les preguntó su nivel de acuerdo con cada afirmación (usando una escala Likert de 10 puntos, con 0 = Sin acuerdo hasta 10 = Total acuerdo) y la clasificación de estándares "mínimos" y "óptimos" para cada recomendación. El número objetivo de encuestados fue de 100 y el nivel de acuerdo establecido fue de 80% para la aceptación

(los niveles más bajos debían ser discutidos por el panel de expertos, con nuevas versiones de las declaraciones propuestas y con una segunda ronda de encuestas si fuera necesario). Una vez que se llegó a un acuerdo, el metodólogo, junto con los becarios clínicos, calificó el nivel de evidencia para cada recomendación en base a los Oxford Levels of Evidence, 2011 (disponibles en http://www.cebm.net/index.aspx?o=5653) y se le asignaron indicadores de calidad relevantes cuando correspondía.

4 RESULTADOS

La necesidad de una transición para adolescentes con enfermedades reumáticas, al igual que para otras enfermedades crónicas, está reconocida por todos los implicados en el proceso; pacientes, padres/cuidadores y profesionales sanitarios que los atienden. Sin embargo, como realizar de forma exitosa esta transición no está del todo claro y se desconocen la organización de este proceso y los recursos empleados en la actualidad en las unidades de reumatología pediátrica en Europa. Por otra parte, la reproductibilidad y efectividad en la práctica clínica de los programas de transición que han sido implementados en la vida real no siempre han sido evaluadas de forma adecuada.

Por este motivo, se realizó un estudio exploratorio acerca de cómo se realiza la transición en adolescentes y jóvenes con enfermedades reumáticas en la práctica clínica real mediante una encuesta a reumatólogos pediátricos europeos. Simultáneamente se realizó una revisión sistemática sobre los programas de transición validados e implementados en la vida real, describiendo los elementos que contienen y prestando especial atención a potenciales recomendaciones, estándares estrategias para su implantación y herramientas para la evaluación de resultados.

En base a estos dos resultados, se llevó a cabo un consenso para disminuir la variabilidad en el manejo de los adolescentes con ERyME durante el proceso de transición y la transferencia a unidades de reumatología de adultos.

Los resultados se presentan en formato de artículos junto con un resumen en español de estos:

Artículo 1: Revisión sistemática y valoración crítica de programas de transición en reumatología / Systematic review and critical appraisal of transitional care programmes in rheumatology (*Semin Arthritis Rheum,* 2016; 35: 1288-1293).

Artículo 2: Atención durante la transición para enfermedades reumáticas en Europa: práctica clínica y recursos disponibles en la actualidad / Transitional care for

rheumatic conditions in Europe: current clinical practice and available resources (*Pediatr Rheumatol Online J*, 2017; 15: 49).

Artículo 3: Normas y recomendaciones para la atención durante la transición de jóvenes con enfermedades reumáticas de inicio en la infancia / EULAR/PReS standards and recommendations for the transitional care of young people with juvenile-onset rheumatic diseases (*Ann Rheum Dis*, 2017; 76: 639-646).

En la siguiente Tabla se expone el factor de impacto y la fecha de publicación de cada uno de los artículos recogidos en la tesis.

Artículo	FI 2016	Cuartil JCR	Decil JCR	DOI	Fecha de publicación
Systematic review and critical appraisal of transitional care programmes in rheumatology	4.498	1	2	10.1016/j.semarthrit.2016.06.003	1/12/2016
Transitional care for rheumatic conditions in Europe: current clinical practice and available resources.	2.283	2	3	10.1186/s12969-017-0179-8	9/6/2017
EULAR/PReS standards and recommendations for the transitional care of young people with juvenile-onset rheumatic diseases.	12.811	1	1	10.1136/annrheumdis-2016-210112	1/4/2017

FI, Factor de impacto
JCR, Journal Citation Report
DOI, Digital Object Identifier

4.1 ARTÍCULO 1 - Programas de transición en enfermedades reumáticas identificados por revisión sistemática.

TÍTULO. Revisión sistemática y valoración crítica de programas de transición en reumatología / Systematic review and critical appraisal of transitional care programmes in rheumatology (*Semin Arthritis Rheum,* 2016; 35: 1288-1293).

OBJETIVO. Identificar, describir y valorar de forma crítica los programas de transición existentes en adolescentes y jóvenes con enfermedades reumáticas.

RESUMEN DE LOS RESULTADOS. Se identificaron y evaluaron 27 artículos, relacionados con 8 programas de transición implementados en 6 países distintos: 4 cubrían todas las ERyME, 3 eran específicos para pacientes con artritis idiopática juvenil y 1 programa era genérico para enfermedades crónicas y se había adaptado para ERyME. Los elementos básicos de estos programas de transición son:

- Un programa de transición por escrito acordado por los equipos multidisciplinares implicados en la unidad pediátrica y de adultos;
- La designación de un coordinador del programa, responsable de que sea llevado a cabo y evaluado de forma apropiada;
- Una planificación individualizada y flexible del proceso, con adaptación al grado de desarrollo del adolescente;
- La educación al paciente y familiares para conseguir la adquisición de conocimientos y el desarrollo de habilidades necesarios para la autogestión de la atención sanitaria;
- Una comunicación efectiva entre las unidades pediátricas y de adultos antes y después de la transferencia del paciente o toma de decisiones compartida en una unidad de transición;
- La transmisión de toda la información relevante a la unidad de adultos, eligiendo el momento más adecuado para trasladar al paciente.

Solo 2 programas de transición de los evaluados proporcionaron evidencia de efectividad de acuerdo con indicadores y medidas de resultado previamente especificadas.

CONCLUSIONES. Esta revisión sistemática resalta la importancia de contar con programas de transición estructurados, cuya implementación puede variar en función de los recursos disponibles a nivel local. Es necesario desarrollar indicadores estandarizados para evaluar las variables de contribuyen al éxito de los programas y su impacto en los adolescentes y jóvenes con enfermedades reumáticas.

SYSTEMATIC REVIEW AND CRITICAL APPRAISAL OF TRANSITIONAL CARE PROGRAMMES IN RHEUMATOLOGY.

Authors: Daniel Clemente, MD[a], Leticia Leon, MS, PhD[b], Helen Foster, MD, PhD[c], Kirsten Minden, MD, PhD[d], Loreto Carmona, MD, PhD[e].

Affiliation: [a]Paediatric Rheumatology Unit, Hospital Infantil Universitario Niño Jesús, Madrid, Spain. [b]Instituto de Investigación Sanitaria del Hospital Clínico San Carlos (IDISSC), Hospital Clínico San Carlos, and Health Sciences, Universidad Camilo José Cela, Madrid, Spain. [c] Musculoskeletal Research Group, Institute Cellular Medicine, Great North Childrens Hospital, Newcastle Hospitals NHS Foundation Trust, Newcastle University, Newcastle upon Tyne, UK. [d] Department of Rheumatology and Clinical Immunology, German Rheumatism Research Centre, and Leibniz Institute, Charité University Medicine Berlin, Berlin, Germany. [e] Instituto de Salud Musculoesquelética, Madrid, Spain.

Corresponding author: Leticia León. Instituto de Investigación Sanitaria del Hospital Clínico San Carlos (IDISSC), Hospital Clínico San Carlos, and Health Sciences, Universidad Camilo José Cela, Madrid, Spain. Email: lleon.hscsc@salud.madrid.org.

ABSTRACT.

Objectives: Identify existing models of transitional care in rheumatic and musculoskeletal diseases (RMD), describe their strengths and weaknesses, and provide support to a consensus initiative to develop recommendations for transitional care.

Methods: A systematic review was conducted to identify publications describing transition programmes in RMD. Eligibility for inclusion required detailed description of the programme. Descriptive information was collected, including country of the programme, target diseases and ages of the patients, resources, elements of the transition process and, when described, outcomes and quality indicators. Quality assessment of the programmes included: level of definition and the evidence base for the programme, availability of quality indicators and evidence of effectiveness.

Results: Overall, 27 articles were identified and evaluated, related to 8 programmes in 6 countries: 4 covered all RMDs, 3 specific for patients with juvenile idiopathic arthritis (JIA) and 1 programme generic for chronic diseases and adapted for RMD. Core elements of these transition programmes included the following: a written transition policy; patient individualized planning and flexibility of transitional care; designation of transition coordinator role; acquisition of knowledge and skills in self- management of care; decision making, shared care and communication between paediatric and adult health care provider teams and a planned transfer to adult rheumatology. Only 2 provided evidence of effectiveness according to previously specified outcome measures.

Conclusions: Transitional care programmes in RMDs are variable in their structures, staffing and processes. There are no standardized measures of outcome or effectiveness. This information provides important valuable insights and strategies to develop transitional care in RMD.

INTRODUCTION.

Transitional care, as defined by the Society for Adolescent Medicine, is "the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centred to adult-oriented health care systems" [1]. Transition is a multidimensional, active process that attends to the medical, psychosocial and educational/vocational needs of adolescents and young adults with special health care needs. In contrast, transfer is the administrative event in which the patient moves from the paediatric or adolescent clinic to the adult office and its medical information pass from one specialist to another [2,3]. A variety of transitional care programmes have been developed to guide patients with chronic diseases and their families through the transition process. Some studies evaluating transition care programmes in other chronic conditions, mainly diabetes, has been published in the last years, but there is a lack of evidence regarding standardized practices in transition care [4,5].

Transition is especially important in youth with juvenile onset rheumatic and musculoskeletal diseases (jRMDs) as approximately 50 % will continue to have ongoing active disease. Many patients will require treatment or have significant sequelae of their rheumatic disease into adult life [6] and there is evidence of worse physical and psychosocial functioning in adults with jRMDs [7]. Although transitional care programmes have been described, their reproducibility and effective- ness in clinical practice remain unclear.

There is an urgent need to move the agenda forward to implement evidence-based transitional care into practice and identifies effective measures of transitional care and quality indicators to measure success [3]. With this in mind, the objective of this review was to identify, describe and critically appraise the existing transitional care programmes in jRMDs.

METHODS

As a part of a European League Against Rheumatism (EULAR) / Paediatric Rheumatology European Society (PRES) working group, a systematic review was performed to select studies that contained validated transitional care programmes, from paediatric to adult healthcare. Once selected, articles were analysed using standard methods for systematic review (http://www.prisma-statement.org/). The information obtained was used to support a consensus initiative to define potential standards and best practice recommendations for transitional care.

Search strategy

The searches were carried out by LL and LC in Medline, Embase and PsycINFO (all from inception until October 2014). These included free text and MeSH terms synonyms of "RMD", "transitional care", and "outcomes" (See Appendix for full search). The search was limited to adolescents, children and young adults and to studies published in English, German and Spanish. In addition, we scanned the reference lists and the "Related articles" function on PubMed (www.pubmed.gov) of the included articles, and searched the abstracts of the ACR, EULAR and PRES meetings available on their official websites.

Study selection

Studies were selected if they described valid transition programmes in jRMDs. The term valid referred to programmes that had been described in sufficient details as to be reproducible and had been implemented and tested in real life conditions). Two reviewers (DC, LL), who had not participated in any of the transition programs published and thus had no conflicts of interest, independently examined the results captured by the searches for articles potentially relevant; first by scanning the title and abstract, and then by detailed review of the full text. Those articles that fulfilled the inclusion criteria were critically and independently appraised by the reviewers, extracting the following information on the transition programmes: country and setting of the programme,

diseases and ages of the patients, resources—staff, transition coordinator, office—, transition process, available indicators, and effectiveness results. Any discrepancies were resolved by consensus with a methodologist (LC).

Quality assessment

In the absence of an available appropriate tool for systematic reviews of health services, we developed an ad hoc scale, based on the work of Campbell [8], to grade the quality of the programmes based on the following dimensions: level of definition / reproducibility, whether the programme was based on previous evidence based benchmark-research, availability of quality indicators, and results/evidence of effectiveness of the programme (Figure 1). Quality was assessed independently and then discussed to reach a consensus, by the reviewers and the methodologist.

RESULTS

The electronic search yielded 521 articles (404 from PubMed, 116 from Embase, and 1 from PsycInfo), plus 6 abstracts from congresses. In addition, a total of 10 references were obtained by reviewing the references of the included articles. The selection by title and abstract left 52 articles plus 6 abstracts for detailed review. Only 26 articles remained for critical appraisal, after excluding duplicated publications and articles not describing or signposting to a programme; these articles corresponded to 8 transition programmes. Another article, presenting results of the evaluation of one of these programmes, was available after the search was finished. Figure 2 shows the flow chart to describe the literature search process.

Tables 1 and 2 describe the programmes identified by the search process and the analysis to focus on the following areas.

Diseases and age of inclusion

All jRMDs were considered in 4 programmes, 3 of them were only applied to patients with juvenile idiopathic arthritis (JIA) and there was one generic programme for patients with various diseases, including a specific adaptation for RMD. Age at inclusion to the transition programme varied, starting as early as 11 years old and not later than age of 17 years. The patient's final transfer to an adult rheumatologist was variable and up to the age of 25 years.

Staff and setting

All the transition programmes began in the paediatric setting and continued into adult health care, sometimes with the development of transition clinics specifically designed for adolescents and young adults with RMD. Transitional care teams were composed of paediatric and adult rheumatologists with interest and training in adolescent rheumatology plus a clinical nurse specialist(s) as a minimum. Other health care professionals that could be part of the transition teams included physiotherapists, occupational therapists, psychologists, social workers and adolescent medicine specialists. Participation of the general practitioner was advocated not only for routine care—specific advice on contraception or intercurrent illnesses- but also for emotional support to patients and family [10,29–31]. All the programmes identified had a transition coordinator, namely a health care professional who was responsible to guide and support young persons and their families during the transition process. In most cases the transition coordinator was a clinical nurse specialist.

Transition processes

All transitional care programmes had a written transition policy and protocol agreed by the paediatric and adult teams that described in detail the transition process. Two components of these programmes were of particular importance: 1) Information and education of both young persons and parents, and 2) Building-up of and promoting skills like self-management and independent living. Shared information included not

only knowledge about RMD and therapies but about generic health, social issues and information about health services (e.g. role of rheumatology professionals, or specificities of paediatric and adult care). In addition to books, booklets or flyers, educational resources included websites and social media. The HEADSS schema was useful to guide consultation to cover all topics pertinent to young people [10,29,31]; this refers to family relationships (Home), level of education and/or expectations of future (Education/Employment), interest, friendship and aspirations (Activities outside school and home, Affect, Ambition), smoking, alcohol, recreational drug use (Drugs), driving (Driving) and diet or weight management (Diet), and sexuality and sexual health (Sex), Suicidality and depression, Sleep. Promoted skills during the transition process included health care autonomy, communication with the clinical team, shared decision making, creative problem solving, resilience and assertiveness. Independent visits of adolescents, separate from their parents, were gradually instituted in the paediatric setting to facilitate acquisition of these skills towards being independent and autonomous. Parents and families were properly informed of the transition process and could discuss all relevant issues with the medical team; they were also encouraged to support their young people and let them take responsibilities in their health care. Peer support groups were established with the objective to share information and to socialize with other teenagers and young adults who understand what is meant to have a chronic disease [9,10,14,28,29,31].

According to all programmes, a transition coordinator was essential for implementing an individualized transition plan tailored to the patient´s social and cognitive maturity and needs; as noted earlier, a nurse specialist usually functioned as transition coordinator, but the role could be performed by other allied health professionals with appropriate training and working in partnership with the clinical team.

Shared care of patients between paediatric and adult team before and after transfer to the adult team was found in most of the programmes. There were, however, variations to the shared care model. In some programmes, members of the adult rheumatology team participated in the paediatric clinics and/or paediatric

rheumatology team members attended clinics in the adult setting [9,10,14,28]. Alternatively, specific transition clinics were developed with both paediatricians and adult rheumatologists in the same setting [29,31].

Transfer of patients to the adult rheumatology clinic took place at a specific age (16-18 years) [9,10,14] or was individualized, occurring when patients met certain "readiness" criteria [28,29,31,34]. These readiness criteria included not only adequate understanding of the disease, treatments or skills acquisition, but also medical aspects, such as stable disease. However, if the patient has not reached the transitional goals at 23 years, continuing in a programme was not considered to be of any further benefit and care was transferred [26]. All relevant information (diagnosis, history, treatment, progress, social and educational arrangements) was usually transferred to the adult team. A "welcome letter" attached to the first appointment letter in the adult clinic was advocated in the MAGICC programme to mark the young patient the new phase of care [9]. Transfer is complete when patients continue follow-up in the adult clinic.

Indicators/results

There was a lack of specified outcome measures in the evaluated transition programmes, with the exceptions of the *Growing up and moving on* program and the *DON'T RETARD* project (Table 3). The 'Growing up and moving on' programme achieved improvements in health-related quality of life, arthritis-related knowledge and satisfaction with rheumatology care in both adolescents and parents, and in vocational readiness markers, although it was unknown whether these improvements were maintained long-term [25,28]. The components of this programme were deemed to be acceptable and useful to adolescents, parents and the rheumatology health professionals [24]. There were also significant improvements in documentation of transitional issues, disease specific educational needs, aspects of adolescent readiness (decision making, communication skills, self-care, independent visits) and several parental needs to promote young people develop their independence [26]. The DON'T RETARD project achieved an improvement in physical, psychosocial and disease–specific health status of adolescents and young adults with an improvement in their quality of

life. The transition programme also influenced and fostered certain parenting behaviours to promote autonomy and independence of young people. However, no differences in illness-related knowledge and medication adherence were observed between the control and the intervention adolescent group [15]. Implementation of the transition clinic proposed by Ammerlaan [9] and the MAGICC [10] programme are under evaluation. Benefits with other programmes are also described, such as improved follow up rates of the patients transferred [28,33] or less "no shows" visits [33].

Quality of the transition programmes

Figure 1 shows the quality of transition programmes using our *ad hoc* scale.

DISCUSSION

We have performed a systematic review on models of transitional care in RMD, comparing and discussing the methodology and outcomes of different programmes around the world that have been implemented and tested in a real-life situation. We have attempted an objective evaluation of the transition programmes in order to determine their quality. The results of this review will serve as reference to generate recommendations for implementation of transitional care models and which can be applied in other settings with flexibility for modifications based on available resources.

We have found several common core features amongst the evaluated programmes. Firstly, a written transition policy, that incorporates the views of the paediatric and adult team, as well as the wider multidisciplinary team as appropriate. Individualized planning and flexibility in timing and duration of the transition process are also key features, acknowledging the heterogeneity in adolescent development and especially when influenced by chronic diseases. Although the age of inclusion in transition programmes is variable, early entry is advisable given that benefits of a transition programme intervention are observed when starting as soon as 11 years old [24,26]. A challenging area to transitional care delivery is to ensure that it is

developmentally appropriate, as the transitional care needs in early adolescence are very different to those in late adolescence.

Another key element is the transition coordinator who, together with the patient and family, takes responsibility for the transition process. Information and education of both young patients and parents along with skills development are imparted to young people to promote disease self-management and self-advocacy in order to allow the patient to negotiate his or her own health care and independently navigate within health-care systems. The youth perceptions of autonomy support from their rheumatologist have been significant in the transition of young people with JIA [50]. Shared care of the patients and communication between paediatric and adult teams before and after transfer facilitates continuity of care during the transition process. A Delphi study found that "assuring a good coordination between paediatric and adult professionals" is the key element and "patient not lost to follow-up was considered the most important indicator" [51]. Finally, it is essential to ensure successful transfers by passing on relevant information to the adult team and having a flexible timing of transfer, ideally when disease activity is relative stable, and the transition team perceive that the patient is "ready".

This literature review has a number of strengths and weaknesses. The strengths centre around the extensive nature of the search conducted, thereby providing a complete and up-to-date overview of systematic models of care for the transition in RMD. Results of the DON´T RETARD programme were published during the preparation of the systematic review and after the search was performed but are included as they are relevant. The main limitations of this review are related to the lack of standardized indicators and outcome measures. The number of structured transitional programmes published in the field of RMDs is limited compared to other chronic diseases (e.g. cystic fibrosis) [52–54]; however, they share many of the same principles. We are aware that other transition clinics may exist but their inclusion did not meet our selection criteria. Also, the fact that very few programmes include evaluation tools, with the exception of the 'Growing up and moving on' and the 'DON´T RETARD' project, make it impossible to judge or to compare their effectiveness. This is similar to that found in other studies of

transition in other chronic diseases, where the need for validated measures of transition readiness and transfer satisfaction are highlighted [55].

Another limitation of the study is the possible emergence of transitional programs that have not been captured by the literature search. This review is part of a EULAR/PRES initiative to develop recommendations and standards for transitional care for young people with jRMDs that started in 2014. Alerts were created in search databases in order to add articles published after our initial search date; this permitted the inclusion of an article [15], but we cannot guarantee the existence of other programs that were not captured.

In summary, this review highlights the importance of structured transition programmes in RMD as well as the importance of developing standardized indicators to evaluate their success. Implementation of transition programmes to improve the transition process needs flexibility and is influenced by the available resources at a local level.

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CONFLICT OF INTEREST STATEMENT

All authors declare no conflicts of interest.

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LIST OF ABBREVIATIONS:

Rheumatic and musculoskeletal diseases (RMD)

Juvenile onset rheumatic and musculoskeletal diseases (jRMDs)

Juvenile idiopathic arthritis (JIA)

Moving on in Adolescence; Growing up In Collaboration and Coping (MAGICC)

Devices for the OptimisatioN of TRansfEr and Transition of Adolescents with Rheumatic Disorders (DON´T RETARD)

Young Adult with Rheumatic Diseases (YARD)

Berlin Transition Programme (BTP)

Paediatric Rheumatology European Society (PReS)

European League Against Rheumatism (EULAR)

Authors contributions:

Daniel Clemente: Manuscript preparation, data collection, methods

Leticia Leon: Manuscript preparation, data collection, methods

Loreto Carmona: Manuscript preparation, methods, quality assessment

Helen Foster: Manuscript preparation, methods

Kirsten Minden: Manuscript preparation, methods

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Table 1. Characteristics of the included studies for transitional care in rheumatology.

Programme	First author and year	Country	Setting	Diseases	Ages (y)	Transition coordinator	Transition team staff			
							Paed Rheum	Adult Rheum	Nurse	Other
Transition clinic	Ammerlaan, 2013[9]	Netherlands	University Medical Centre Utrecht	JIA	16 to 25	Nurse specialist	✓	✓	✓	Not specified
MAGICC	Tattersall,2012[10]	United Kingdom	Sheffield Hospitals	Childhood onset RMD	11 to 25	Nurse specialist	✓	✓	✓	Physiotherapist, Occupational therapist
DON'T RETARD	Previous research: Eyckmans 2011[11], Hilderson 2012[12], 2013a[13] Description: Hilderson 2013b[14] Results: Hilderson 2015[15]	Belgium	University Hospitals Leuven	JIA	14 to 16	Yes**	✓	✓	✓	Physiotherapist, Psychologist
Growing up and moving on	Previous research: Robertson 2003[16], Shaw 2004a[17], 2004b[18], 2004c[19], 2007a[20], McDonagh 2004[21], 2006a[22] Description: Shaw 2005[23], McDonagh 2006b[24] Results: McDonagh 2006b[24], 2007[25], Robertson 2006[26],Shaw 2007b[27]	United Kingdom	10 centres across UK	JIA	11 to 17	Yes***	✓	✓	✓	Variable depending on centre but included physiotherapist and occupational therapist
Rheumatology transition programme	Rettig 1991[28]	United States of America	Children´s Hospital of Philadelphia/Hospital of the University of Pennsylvania	Childhood onset RD	17 to 19	Nurse specialist	✓	✓	✓	Social worker
YARD clinic (Vancouver)	Tucker 2005 [2007][29], 2008[30]	Canada	British Columbia Children´s Hospital of Vancouver	Childhood onset RD	14 to 23	Nurse specialist	✓	✓	✓	Physiotherapist, Occupational therapist, Adolescent Medicine specialist
YARD clinic (Calgary)	Description: Miettunen 2008[31], Guelber 2008[32] Results: Labrie 2008[33]	Canada	Alberta Children´s Hospital Calgary Foothills Hospital in Calgary, Alberta	Childhood onset RD	14 to 25	Nurse specialist	✓	✓	✓	Social worker, Physiotherapist
Berlin Transition Programme (BTP)	Minden 2014[34]	Germany	Some regions of Germany	Childhood onset RD*	16 to 20	Case manager	✓	✓	✓	Not specified

*Generic programme for patients with various diseases, including a specific adaptation for RMD

** and ***Health professional involved as transition coordinator not specified

Table 2. Characteristics of the included studies for transitional care in rheumatology – continued.

Programme	Office			Individualized transition plan				Shared care	Age of transfer
	Paediatrc clinic	YARD clinic	Adult clinic	Written transition policy	Education	Skills development	Communication and tools		
Transition clinic	✓		✓	Yes	Yes	Yes	Digital Rheuma Portal (online portal for e-consultation) Website: http://www.jong-en-reuma.nl Training programme: http://www.reuma-uitgedaagd.nl Peer support group	Yes	At 18 y
MAGICC	✓	✓		Yes	Yes	Yes	Website:http://www.sheffieldchildrens.nhs.uk/patients-and-parents/transition.htm Peer support group	Yes	At 16 y Transfer proforma
DON'T RETARD	✓		✓	Yes	Yes	Yes	Helpline (transition coordinator) Website: http://www.kuleuven.be/switch2/rheuma.html Peer support group	Yes	At 16 y
Growing up and moving on	✓		✓	Yes	Yes	Yes	Guided by local program coordinator Resources provided: Individualized transition plan templates Rheumatology Adolescent Planner (RAP) Filofax Rheumatology Adolescent Planner (RAP) resource books for parents and for local programme coordinators Adolescent rheumatology resource directories for health professionals Website: http://www.arthritiscare.org.uk	No	At 16-18 y Variable between centres
Rheumatology transition programme	✓		✓	Yes	Yes	Yes	Guided by transition coordinator	Yes	Individualized between 17 to 19 y after readiness to transfer is checked
YARD clinic (Vancouver)	✓	✓		Yes	Yes	Yes	Guided by transition coordinator	Yes	Individualized between 20 to 23 y Readiness to transfer checklist
YARD clinic (Calgary)	✓	✓		Yes	Yes	Yes	Guided by transition coordinator Peer support group	Yes	Individualized, after readiness to transfer is checked
Berlin Transition Programme (BTP)	✓		✓	Yes	Yes	Yes	Carried by case manager Resources provided: Information booklet for physicians and patients/families, questionnaires, Checklist, T (transition)- Booklet, flyer	No	Individualized, when transition programme of 2 y is finished

Table 3. Characteristics of the included studies for transitional care in rheumatology (Third part: Indicators and evidence of performance).

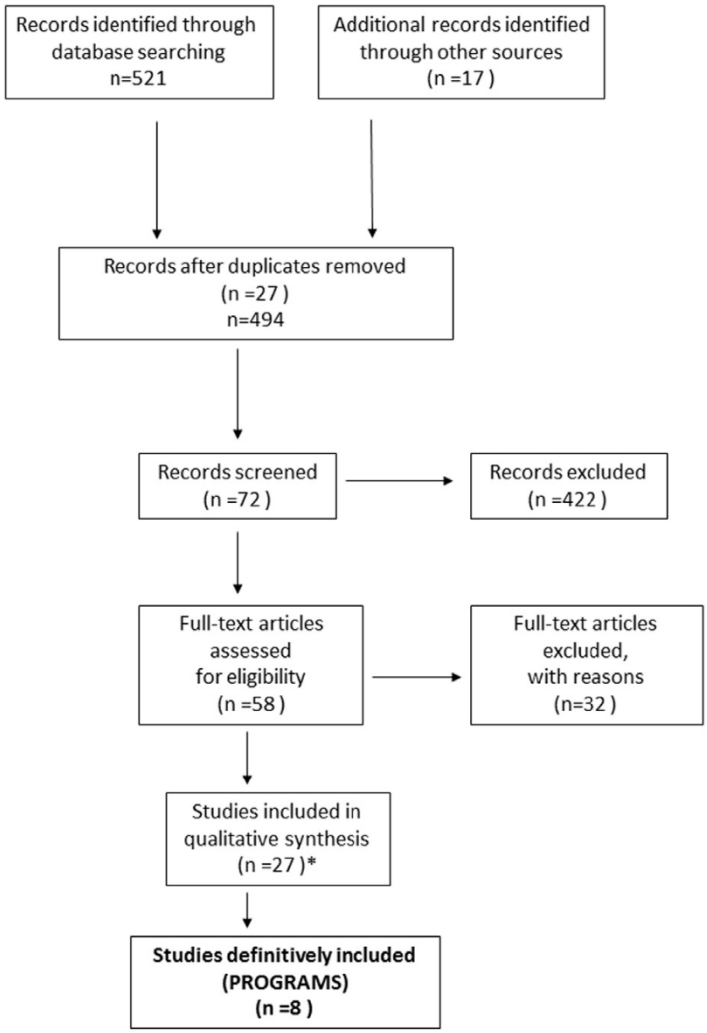
Transition clinic	Not specified	A trial in progress. No results yet
MAGICC	Not specified	No results
DON'T RETARD	Primary outcome: Self-perceived health status of the patient: Paediatric Quality of Life Inventory (PedsQL), generic (PedsQL 4.0 Generic Core Scale) and disease specific module (PedsQL 3.0 Rheumatology Module) [35-37]	Improvement in physical, psychosocial and disease-specific health status
	Secondary outcomes: Patients´ health status as perceived by the parents:PedsQL 4.0 Generic Core Scale and PedsQL 3.0 Rheumatology Module	No effect or small negative effect in physical, psychosocial and rheumatic specific health status
	Patient´ self-reported secondary outcomes: Medication adherence: Visual Analogue Scale (VAS) and SWISS HIV Cohort Study Adherence Questionnaire (SHCS-AQ) [38] Illness-related knowledge: The Modified Patient Knowledge Questionnaire (PKQ) [39] Global quality of Life: Linear Analogue Scale (LAS) [40] Fatigue: Multidimensional Fatigue Inventory (MFI-20) [41]	 No effect on medical adherence Increase in Illness knowledge similar to control group Improvement in quality of life Small negative effect in reduction in general fatigue
	Parenting Dimensions[42-44] Promotion of Independence by parents: Promotion Independence Scale (PI) Support of autonomy: Autonomy Support Scale (PVF) Behavioural control: Parental Regulation Scale (PRS-YSR revised to parent self-report). Psychological control: Psychological Control Scale (PCS-YSR revised to parent self-report)	 Increase in autonomy support and promotion of independence, with a reduction in behavioural control No beneficial impact on psychological control
	Disease parameters: Clinical status of disease activity and clinical remission on/off medication: preliminary criteria of Wallace et al. [45] Functional status: patient-reported and parent-reported Childhood Health Assessment Questionnaire (CHAQ-DI) [46-48]	 No results
Growing up and moving on	Primary outcome: Health-related quality of life (HRQL): Juvenile Arthritis Quality of Life Questionnaire (JAQQ)[49]	Improvement of health-related quality of life
	Secondary outcome measures: Arthritis-related knowledge: 16-item disease-specific multidimensional measure with multiple choice response format	 Improvement of disease knowledge for adolescent and parents
	Satisfaction with rheumatology care: patient and parent satisfaction were measured using 22- and 27-item measures respectively	Improvement of patients and parents satisfaction
	Independent health behaviours (self-medication and independent consultations): closed questions	No significant changes in self-medication or independent consultations were observed at either the 6 or the 12 month assessments
	Pre-vocational experience (household chores, work experience, career advice and career aspirations): series of closed and open questions	Improvement of vocational readiness markers
Rheumatology transition programme	Not specified	Improvement of adherence to appointments after transfer to adult rheumatology services
YARD clinic (Vancouver)	Not specified	No results
YARD clinic (Calgary)	Not specified	Improvement of adherence to appointments Independent visits implemented Good control of the disease during transition Improvement vocational readiness All patients received allied health care support

Figure 1. Quality grading of programmes.

	Definition (D)	Research-based program (RP)	Quality indicators (QI)	Results / Evidence of effectiveness (R)
Transition clinic				
MAGGIC				
DON´T RETARD				
Growing up and moving on				
Rheumatology transition programme				
YARD Clinic Vancouver				
YARD Clinic Calgary				
BTP				
	 The program is sufficiently described and defined as to make it reproducible  The program is described but some aspects of the description are too open or ambiguous  In general, the program is not described or it is too open or ambiguous	 The program design was based on extensive previous research (Focus groups, Delphi, surveys, systematic review, audits...)  The program design was based on discussion group + some other type of qualitative research  There is no indication as to how the program was designed	 The program defines quality indicators to measure success The program does not define quality indicators but they are easy to develop based on the recommendations The program does not define quality indicators and it is not easy to develop QI based on the recommendations	At least an intervention trial shows the effectiveness of the program There is some evidence that the program has improved outcomes (health, resource consuming...) in the area There is no evidence that the program has improved the health outcomes or any other evidence or results.

MAGGIC: Moving on in Adolescence, Growing up In Colaboration and Coping; DON´T RETARD: Devices for OptimisatioN of TRansfEr and Transition of Adolescents with Rheumatic Disorders; YARD: Young Adults with Rheumatic Diseases; BTP: Berlin Transition Programme

Figure 2. Flow chart of the selection of the studies.



*One article, presenting results of the evaluation of one of these programmes, was available after the search was finished

(Supplementary material) All terms were search in MeSH + All fields

Population	**AND**	**OR**	adolescents adolescence children adolescents young adults young age childhood children pediatric patients Paediatric youth	young adults infancy children teen teenagers youngsters juvenile preadult young
		OR	"Arthritis, Juvenile"[Mesh] Juvenile Arthritis Arthritis, Juvenile Idiopathic Idiopathic Arthritis, Juvenile Arthritis, Juvenile Rheumatoid Rheumatoid Arthritis, Juvenile Enthesitis-Related Arthritis, Juvenile Arthritis, Juvenile Enthesitis-Related Enthesitis Related Arthritis, Juvenile Juvenile Enthesitis-Related Arthritis Juvenile Chronic Arthritis Juvenile Idiopathic Arthritis Oligoarthritis, Juvenile Juvenile Oligoarthritis Polyarthritis, Juvenile, Rheumatoid FactNegative Polyarthritis, Juvenile, Rheumatoid FactPositive Psoriatic Arthritis, Juvenile Arthritis, Juvenile Psoriatic Juvenile Psoriatic Arthritis Systemic Arthritis, Juvenile Arthritis, Juvenile Systemic Juvenile Systemic Arthritis Arthritis, Juvenile Chronic Chronic Arthritis, Juvenile Juvenile Rheumatoid Arthritis Juvenile-Onset Still Disease Juvenile Onset Still Disease Still Disease, Juvenile-Onset Still Disease, Juvenile Onset Still's Disease, Juvenile-Onset Juvenile-Onset Still's Disease Still's Disease, Juvenile Onset Juvenile-Onset Stills Disease Juvenile Onset Stills Disease Stills Disease, Juvenile-Onset "Lupus Erythematosus, Systemic"[Mesh] Localized Scleroderma Sclerodermas, Localized Scleroderma, Circumscribed Circumscribed Scleroderma Dermatosclerosis Morphea Morpheas Scleroderma, Linear Linear Scleroderma Frontal Linear Scleroderma en Coup de Sabre “Scleroderma, Systemic"[Mesh] “Vasculitis"[Mesh] “Dermatomyositis"[Mesh] Dermatomyositides	

			Dermatopolymyositis Dermatopolymyositides Polymyositis-Dermatomyositis Polymyositis Dermatomyositis Polymyositis-Dermatomyositides Dermatomyositis, Childhood Type Childhood Type Dermatomyositides Childhood Type Dermatomyositis Dermatomyositides, Childhood Type Dermatomyositis, Adult Type Adult Type Dermatomyositides Adult Type Dermatomyositis Dermatomyositides, Adult Type
Intervention	**OR**		Transition care "Health Transition"[Mesh] "Transition to Adult Care"[Mesh] Transference of care Transition to adult care Pediatric Transition To Adult Care transition clinics transition care transfer of care transfer transition counseling transition care health-care delivery transition to adulthood transition agenda Continuing patient care Pilot study Coordinated care Readiness Transitioning Step Management Preparing
Outcome	**OR**		Transition unit characteristics Standards Standard procedure Key Recommendations Quality indicators Tools Strategies Programs Guide Guidelines Guidance Evidence Efficacy Criteria Criterion Protocol Indicator Indication Draft Models Skills Scheme Plan Approach Method Project Rule

4.2 ARTÍCULO 2 - Situación de la transición para las ERyME juveniles en Europa.

TÍTULO. Atención durante la transición para enfermedades reumáticas en Europa: Práctica clínica y recursos disponibles en la actualidad / Transitional care for rheumatic conditions in Europe: current clinical practice and available resources (*Pediatr Rheumatol Online J,* 2017; 15: 49).

OBJETIVO. Valorar el manejo actual de la transición de los reumatólogos pediátricos en Europa y los recursos disponibles antes de la difusión de las recomendaciones para la transición de EULAR /PReS.

RESUMEN DE LOS RESULTADOS. Se envió una encuesta a 276 reumatólogos pediátricos de distintos centros europeos de PRINTO, con una tasa de respuesta del 44%, obteniéndose representación de 115 centros de 22 países. Aunque 32/121 (26%) respondieron que sus centros no ofrecían servicios de transición, la mayoría (99%) consideraba necesario proporcionar una atención específica en la transición a la atención en las unidades de adultos.

Una minoría (<30%) de los encuestados desarrollaba un programa de transición estructurado por escrito, siendo más frecuente desarrollar un proceso de transición informal (46%). El personal designado para llevar a cabo la transición solo estaba disponible en una minoría de centros. En algunos casos se ofrecía una verdadera atención multidisciplinar, implicando además de a médicos, a enfermeros (35%), fisioterapeutas (15%), psicólogos (15%), trabajadores sociales (8%) y terapeutas ocupacionales (2%). La existencia de un miembro del equipo responsable de coordinar la transición fue reconocida en muchos centros (64% de los encuestados).

Aunque los conocimientos y las habilidades que se consideran importantes para que los adolescentes y jóvenes puedan ser autónomos a la hora de integrarse en las unidades de adultos sean similares en los centros encuestados, solo un 36% utiliza una lista de verificación de la adquisición de éstos como parte de la atención individualizada de transición.

CONCLUSIONES. Esta encuesta demuestra limitaciones en la atención transicional de adolescentes y jóvenes con ERyME y una escasa disponibilidad de recursos. A pesar de ello, existe un fuerte compromiso de reumatólogos pediátricos y adultos para mejorar la atención en el proceso de transición, siendo las recomendaciones EULAR/PReS útiles y oportunas para favorecer los cambios necesarios en la práctica clínica.

TRANSITIONAL CARE FOR RHEUMATIC CONDITIONS IN EUROPE: CURRENT CLINICAL PRACTICE AVAILABLE RESOURCES.

Authors: Daniel Clemente, MD[a], Leticia Leon, MS, PhD[b], Helen Foster, MD, PhD[c], , Loreto Carmona, MD, PhD[d], Kirsten Minden, MD, PhD[e].

Affiliation: [a]Paediatric Rheumatology Unit, Hospital Infantil Universitario Niño Jesús, Madrid, Spain. [b]Instituto de Investigación Sanitaria del Hospital Clínico San Carlos (IDISSC), Hospital Clínico San Carlos, and Health Sciences, Universidad Camilo José Cela, Madrid, Spain. [c] Musculoskeletal Research Group, Institute Cellular Medicine, Great North Childrens Hospital, Newcastle Hospitals NHS Foundation Trust, Newcastle University, Newcastle upon Tyne, UK. [d] Instituto de Salud Musculoesquelética, Madrid, Spain. [e] Department of Rheumatology and Clinical Immunology, German Rheumatism Research Centre, and Leibniz Institute, Charité University Medicine Berlin, Berlin, Germany.

Corresponding author: Kirsten Minden. Department of Rheumatology and Clinical Immunology, German Rheumatism Research Centre, and Leibniz Institute, Charité University Medicine Berlin, Berlin, Germany. Email: minden@drfz.de.

ABSTRACT

Objective. To assess European pediatric rheumatology providers' current clinical practices and resources used in the transition from child-centered to adult-oriented care.

Methods. European pediatric rheumatologists were invited to complete a 17-item anonymised e-survey assessing current transition practices, transition policy awareness, and needs in advance of the publication of PRES/EULAR recommendations on transition.

Results. The response rate was 121/276 (44%), including responses from 115 centers in 22 European Union countries. Although 32/121 (26%) responded that their centers did not offer transition services, the majority (99%) agreed that a formalized process in transitioning patients to adult care is necessary. A minority (<30%) of respondents stated that they have a written transition policy although 46% have an informal transition process. Designated staff to support transitional care were available in a minority of centers: nurse (35%), physiotherapist (15%), psychologist (15%), social worker (8%), and occupational therapist (2%). The existence of a designated team member to coordinate transition was acknowledged in many centers (64% of respondents) although just 36% use a checklist for young people as part of individualized transitional care.

Conclusion. This survey of European pediatric rheumatology providers regarding transitional care practices demonstrates agreement that transitional care is important, and wide variation in current provision of transition services exists.

INTRODUCTION

Juvenile onset rheumatic and musculoskeletal diseases (jRMDs) have high impact on all aspects of the lives of children, young people (YP) and their families. Based on prevalence rates, it is estimated that there are 75,000 children under 16 years old with juvenile idiopathic arthritis (JIA) [1] and about 2,500 with systemic lupus erythematosus (SLE) in Europe [2]. These conditions persist into adulthood for many individuals, with continuing disease activity and need for ongoing medication with potential disease-associated morbidity, life-long disability and psychosocial impact [3-5]. During the period of transition, YP with jRMDs have to cope with tremendous physical, emotional and social changes and also with important change in the delivery of clinical care as they move into adult services. Providing transitional care services during this period is necessary to ensure that YP can take control of their health care needs, engage with health care providers and ultimately emerge into adulthood with their optimal function and potential [6-7]. High-quality, developmentally appropriate healthcare service provision requires the involvement of the YP and the family and also a continuous communication between all persons and provider services involved in transitional care [8].

Whilst the rationale and aims of transitional care in young people with jRMD are clear, organization and delivery of transitional care and what constitutes the ideal service, remains unclear [9,10]. Previous surveys in the UK, North America and Europe [11-13] have demonstrated variable provision of care and resources, unmet education and training needs for health care providers, and the need for guidance on the process. This state of play facilitated the drive for the PReS / EULAR working party for transitional care to be set up in 2014 with subsequent published recommendations [14]. The purpose of our survey was to assess as a baseline, the European pediatric rheumatology providers' transition practices and resources in advance of the PReS / EULAR recommendations for transition being widely available.

METHODS

European pediatric rheumatologists were invited to complete an anonymised 17-item e-survey assessing their current transition practice. The questionnaire was developed by the authors (DC, LL, HF, LC, KM) after a systematic literature review and critical appraisal of transitional care programs in rheumatology [10] and two face-to-face meetings of the PReS / EULAR working party for transitional care [14]. Items were included that inquired current transition practices, agreed key elements of transition programs and available resources. For comparability reasons, items 1 to 3 were matched with questions of the North-American CARRA survey by Chira et al. [12]. The survey was distributed by the Coordinating Centre of PRINTO (Pediatric Rheumatology INternational Trials Organisation) via SurveyMonkey© to the PRINTO directors of all PRINTO centers in 25 European countries. PRINTO is an international network of academic, clinical centers actively engaged in the research/clinical care of children and adolescents with pediatric rheumatic diseases. One reminder was sent to increase the response rate. The questionnaire included items about the transitional care service, staffing, process and resources. The e-survey was piloted by the authors to optimize clarity and ease of administration online.

The study was conducted in cooperation with PRINTO. We did not collect personal identifiable information, such as telephone numbers, names of individuals or institutions, or addresses. The survey respondents voluntarily participated in the study giving their consent to data collection for the purpose of scientific research.

Descriptive statistics are presented and where possible the results were compared with the North American 2014 survey [12] using proportions z test statistical analysis, assuming independence of the two cohorts and $\alpha = 0.01$, given the multiple comparisons being made [15,16]. The responses of rheumatology centers from different levels of care were compared by the chi-squared test. The 99% CI of the difference between surveys is provided.

RESULTS

Respondent demographics and characteristics

The link to the survey was e-mailed to pediatric rheumatologists (n=276) from PRINTO centers in 25 European Union countries in April 2016. Of these, 121responded (44% response rate), representing 115 centers in 22 countries, and thus much of the European Union. Exceptions were Bulgaria, Estonia and Lithuania, that did not reply, and Cyprus, Malta and Luxemburg, which do not have PRINTO centers. The 115 centers with response correspond to 88% of the PRINTO centers currently listed on the PRINTO-website in those 22 countries. These represent centers with a special interest in research and possibly also in transition. Most respondents (n=60; 49%) work in a university-affiliated practice, 39% (n=47) in a designated children's hospital, 4% (n=5) in private practice and 7% (n=8) in other settings.

Transition policy, staff involved, tools and resources

Most (n=120; 99%) respondents regarded a transition policy as being necessary for good clinical practice. The majority stated that they follow an informal transitional care with only 1 in 4 centers having a written transition policy (Table 1). Most rheumatology centers (respondents n=88; 73%) regularly offer transition services for YP with rheumatic diseases, with a wide range of health professionals involved including pediatric and adult rheumatologists and nurses being most common (Table 2). There is a designated staff member who has primary responsibility for coordinating transition process in approximately two thirds of centers (respondents n=77, 64%); the staff member is usually a doctor (n=60; 78%) or a nurse specialist (n=14; 18%). A minority (respondents n=9/119; 7.6%) of transition units receive designated funding or reimbursement for their services, with health insurance companies, grants, pharmaceutical companies, or university as sources.

A minority of centers (respondents n=44; 36.4%) use a checklist as part of an individualized transition plan that includes the topics shown in Table 3. The majority of these (40/44; 90.9%), agreed that confirmation of the first contact with adult rheumatologist is critical. Other important topics were the importance of knowledge

about disease and treatment issues (39/44; 88.6%) as is the encouraging self-management and independent visits by YP without their parents (35/44; 79.5%).

Of note, less than 10% of centers (respondents n=11) use a specific readiness for transfer instrument, of these, 5 use a self-developed readiness scale. Other instruments used are the Transition Readiness Assessment Questionnaire (TRAQ), TRANSITION Scale, Ready Steady Go [12].

A minority of respondents (18/121; 15%) reported to use or recommend specific resources for YP and their families. These were mainly websites providing information on juvenile rheumatic diseases, but also on transition aspects, such as the PRINTO website [www.printo.it/pediatric-rheumatology] and the website on transition of the Competence Network Patient Training in Adolescence KomPaS e. V. [www.between-kompas.de]). Furthermore, a minority (38/121; 31%) use information technology to facilitate communication with YP; mainly Short Message Service (SMS), web platforms, email, or apps and the remainder tend to communicate by telephone.

Among the rheumatology centers from different levels of care, some differences regarding transition care were observed. University affiliated practices offered somewhat less often transition services (69% vs. 78%), had less often a written transition policy (22% vs. 41%) and used significantly less frequently a checklist as part of individualized transitional care (23% vs. 48%, p=0.011) than children's hospitals.

Figure 1 is a pragmatic attempt to compare the data from our survey and previous published data from North America [12]. Notably the majority of North American respondents (89%) work in a university-affiliated practice in contrast to 49% of their European colleagues (p<0.01; 99% CI: 0.29, 0.49).Current transitional care services in Europe and North America show significant differences; the presence of a designated staff member who coordinates transition (64% in Europe vs 30% in North America; p<0.01; 99% CI: -0.45, -0.22) and the availability of a written transition policy (26% in Europe vs 8% in North America; p<0.001; 99% CI: -0.26, -0.08).Nevertheless, the percent of informal transition process is similar across surveys (46% vs. 42%; non-significant).

DISCUSSION

This is the first survey to focus on the current practices (rather than solely attitudes or opinions) in transitional care among European pediatric rheumatology providers. As in other chronic diseases that start in childhood, YP with jRMD need specific care during the period of transition from pediatric services to the adult specialty care setting. However, research about current practice and the implementation of transition recommendations are scarce and almost half of the YP with jRMD do not make a successful transfer to adult rheumatology and are, therefore, at increased risk of unfavorable outcomes [5,18,19].

Recently, Eular/PreS recommendations were issued for transitional care throughout Europe [14]. These recommendations highlight the need for a written transition policy, and yet less than a third of respondents in our survey indicated that their department had a formal transition program; an "informal" approach was reported to be more common although further information was not given in the survey. It is hoped that the use of the recommendations and a consistent approach to the transition program is more likely to address the structure, assessments of patient readiness and service outcomes. We note that only a small percentage of transition clinics receive funding or reimbursement for the service and it is possible that one barrier to implement more ambitious transition programs may be insufficient resources. Interestingly, the Eular/PreS recommendations include the need to secure funding for provision of transitional care, in order to avoid some of the major barriers as highlighted by the North American survey and a common shortcoming in Europe [14].

The presence of a transition coordinator is another key element of transition and one of the recommendations; the aim being to ensure implementation and evaluation of the transition program and improving communication between health professionals, YP and families [8,14]. Most respondents of the survey have a designated staff member as transition coordinator; however, we note that the designated staff member is often a doctor rather than a nurse, youth worker or other professional. The Eular/PreS recommendations do not specific the health care professional to be the coordinator but

that the individual is clearly identified within the team and has the appropriate skills to facilitate liaison between the pediatric and the adult care teams [14].

Information and education of YP about their disease, treatments and promotion of independent visits to clinic without parents are ideal components of an individualized transition plan. However, it is important to include systematically other important areas of adolescence health, such as risk behavior, substance use, sports, nutrition or sexuality. The HEADSS assessment (Home, Education/Employment, peer group, Activities, Drugs, Sexuality, Suicide/depression) is useful to guide the interview about all aspects of adolescent life and facilitate communication with YP [20]. However, to address all relevant transition issues, it might be necessary to have a multidisciplinary team (i.e., psychologist to handle mental health or physiotherapist to advice about sports and routine exercises). This approach is clearly recommended in the Eular/PreS document [14]. The survey revealed that currently only a minority of centers offers multidisciplinary transitional care services. Just 35% of the centers have a nurse, 15% a psychologist and 8% a social worker available for addressing transition-relevant issues.

Our survey did not specifically address timing of transfer. However, it should be noted that only 9.5% use a readiness instrument to validate transfer, reflecting the informal approach to transition in the majority of centers. This issue was heavily discussed in the Eular/PreS recommendations, as some countries had a designated age for transfer in all cases and independently of YP readiness. Finally, an age was not specifically recommended for the transfer, although it was proposed that the transition process should start as early as possible [14]. Some strategies to facilitate transfer include previous communication with adult services with relevant information about individual YP or shared clinics between pediatric and adult health care professionals. In this way, timing of transfer could be more flexible, and could be delayed if necessary until the disease is stable and the transition team considered that the patient is “ready”.

Where possible and with the questions that can be compared, we were able to comment on differences between European and North American respondents. European providers more often have a transition coordinator (often a doctor) and are

more likely to use a written transition policy statement. In both surveys, respondents manifest a strong desire for rheumatology-specific guidelines for transition [12] and for these to be implemented across pediatric, adolescent and adult rheumatology health care settings.

In summary, this survey has demonstrated limitations of existing transition practices and paucity of resources. Nonetheless there is a strong commitment within the rheumatology communities (both adult and pediatric), to improve existing transitional care provision. The EULAR/PReS recommendations are therefore timely and important as a much needed catalyst for change within the rheumatology community.

DECLARATIONS

Ethics approval and consent to participate: not applicable.

Consent for publication: not applicable.

Availability of data and material: the data of this survey are available from the last author on reasonable request.

Competing interests: the authors declare that they have no competing interests.

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Authors' contributions: all authors were involved in drafting the article and approved the final version to be published. Survey conception and design: KM and HF. Acquisition of data: KM. Analysis and interpretation of data: DC, LL, HF, LC, KM.

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Table 1. Current transition policies at the respondents' units

Regarding transition policies*	N	%
We have a written transition policy, which we follow most of the time.	29	23.9
We have a written transition policy, but we do not follow it most of the time.	4	3.3
We do not have a written transition policy, but follow a fairly standard, informal procedure in transitioning our patients.	56	46.2
We are working on developing a transition policy, but do not yet have one formalized.	21	17.3
We do not have a transition policy but are interested in developing one.	10	8.2
I do not think that a transition policy is necessary at this point.	1	.8
I have not given it much thought.	0	-

* Percent obtained from 121 affirmative responses.

Table 2. **Staff involved in the transition services of the respondents' units.**

Health professionals in the transition service	N	%
Paediatric rheumatologist	92	76.0
Adult rheumatologist	77	63.6
Nurse	42	34.7
Psychologist	18	14.8
Physiotherapist	18	14.8
Social worker	10	8.2
Occupational therapist	3	2.4
Others*	11	9.0

* Include: internist, medical assistant, expert patients, orthopedic surgeon, gynecologist

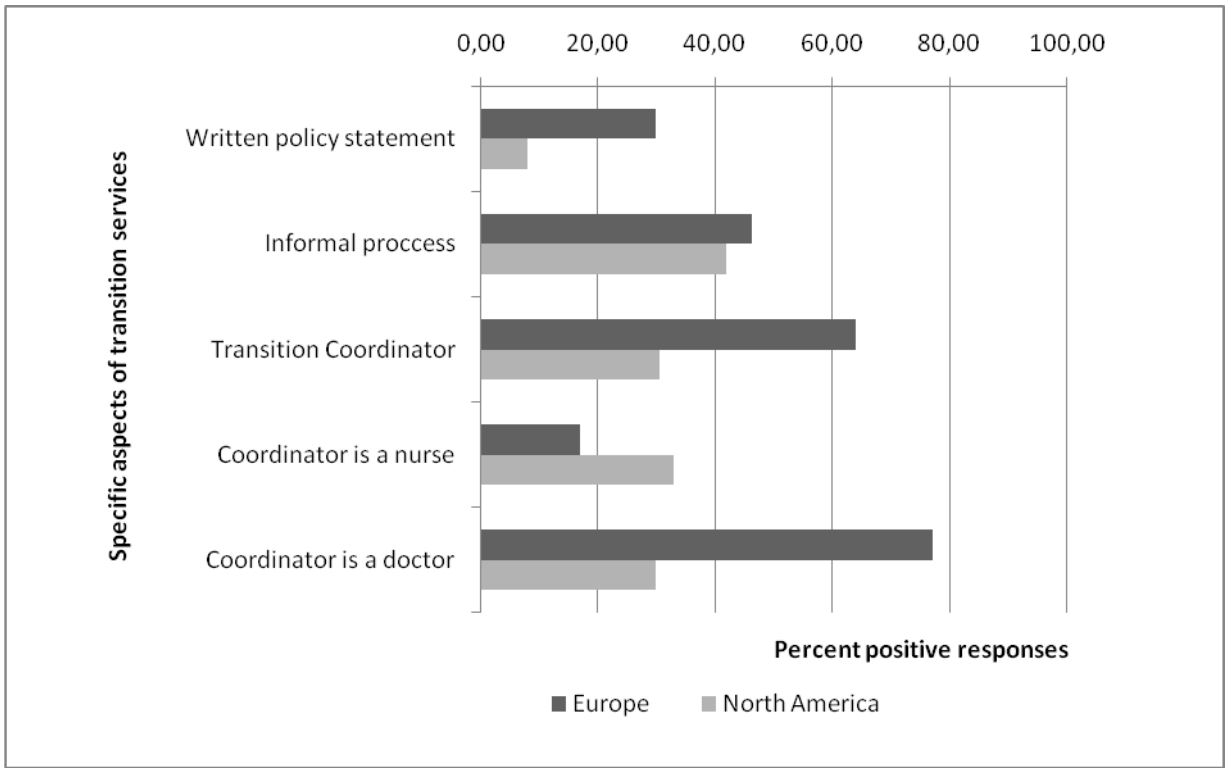
Table 3. Components of existing individualized transition plans.

Checklist components*	N	%
Self-management, patient is seen without parents	35	79.5
Disease and treatment knowledge	39	88.6
Name of disease	39	88.6
Being able to describe disease course	32	72.7
Signs and symptoms of disease flare	33	75.0
Signs and symptoms that require an urgent consultation	30	68.1
Treatment information	40	90.9
Possible side effects of treatment	36	81.8
Health behaviour	31	70.4
Risk behaviour	29	65.9
Alcohol use, smoking, illegal drugs use	35	79.5
Nutrition	27	61.3
Sports	31	70.4
Mental health	27	61.3
Sexuality, contraception and sexual health	34	77.2
Future plans	33	75.0
Educational achievements	30	68.1
Vocational readiness	22	50.0
Knowledge of support resources	28	63.6

Checklist components*	N	%
Mobility, living alone, travel	24	54.5
Medical summary available	34	77.2
Knowledge of differences between pediatric and adult rheumatology care	36	81.8
Knowledge about the health system (health insurance, general practitioner/family doctor, health care specialist)	21	47.7
Transfer readiness	28	63.6
First contact to adult rheumatologist	40	90.9

* Percent obtained from 44 affirmative responses on having a checklist.

Figure 1. Comparison of transitional care services offered by European and Childhood Arthritis and Rheumatology Research Alliance (CARRA) paediatric rheumatology providers.



Appendix 1

Questions regarding transition practices:

1) Regarding transition from pediatric to adult care, select one of the below which best describes your practice:

❍My practice has a written transition policy which we follow most of the time

❍My practice has a written transition policy, but we do not follow it most of the time.

❍We do not have a written transition policy but follow a fairly standard, informal procedure in transitioning our patients

❍We are working on developing a transition policy but do not yet have one formalized

❍We do not have a transition policy but are interested in developing one

❍I do not think that a transition policy is necessary at this point

❍I have not given it much thought

2) Is there a designated staff member in your practice who has primary responsibility for coordinating transition process?

❍Yes ❍ No

if yes (you do have a designated staff member in your practice), what is his/her role?

❍Nurse

❍Office manager/assistant

❍Social worker

❍Physician

❍Other, please specify ……

3) Does your institution regularly offer transition services for adolescents with rheumatic diseases?

❍Yes ❍No

if yes: Who is usually involved in the transition service (please indicate all that apply)

❍pediatric rheumatologist

❍adult rheumatologist
❍nurse
❍psychologist

❍ physiotherapist
❍ social worker
❍ occupational therapist
❍ other persons, namely …….. please specifiy

Do you receive funding or reimbursement for the service?

❍Yes ❍ No

if yes: from whom? ❍ government

❍ health insurance company

❍ others, namely …….. please specifiy

4) Does your institution use a checklist for providing individualized transitional care?
❍Yes ❍No

if yes: which aspects are addressed by the checklist (please tick the appropriate)

❍ self-management, patient is seen without parents

❍ disease and its treatment ❍name of disease
- ❍being able to describe disease course
- ❍ signs and symptoms of disease flare
- ❍ signs and symptoms that require an urgent consultation
- ❍kind of treatment
- ❍ possible side effects of treatment

❍ health behavior

❍ risk behavior
- ❍alcohol use, smoking, illegal drug use
- ❍ nutrition

❍sports
❍ sexuality, anticonception

❍ future plans, perspectives
- ❍educational achievements
- ❍vocational preparation and training

❍ knowledge of available ressources, legal support packages

- ❍ mobility, living alone, travel

❍ having a medical summary
❍ knowing the differences between pediatric and adult rheumatology care
❍ knowledge about the health system (health insurance, general practitioner/family doctor; health care specialist)
❍ transition readiness
❍ first contact to adult rheumatologist
❍ others, namely …… please specifie

5) Does your institution use a specific validated readiness instrument?
❍Yes ❍No
if yes: please report which one

❍ Transition Readiness Assessment Questionnaire (TRAQ)
❍ Am I ON TRAC for Adult Care
❍ Transition-Q
❍ Self-Management Skills Assessment Guide (SMSAG)
❍ Readiness for Adult CarE in Rheumatology (RACER)
❍ TRxANSITION Scale
❍another, namely …….. please specifiy

6) Does your institution use or recommend specific resources (e.g. specific websites like http://www.sickkids.ca/Good2Go/for-health-care-providers/Index.html or www.transitioninfonetwork.org.uk, brochures)?

❍Yes ❍No
if yes: please report which one: ……..

7) Do you use new media for the communication with adolescent patients?
❍Yes ❍No
if yes, which one

❍ SMS
❍ web platform
❍ Apps
❍ social media
❍ others, namely ……..please specifiy

Many thanks for accepting to answer this questionnaire.

Please answer finally these questions.

Please select the one response which best describes your practice

- ❍University affiliated practice
- ❍General Children´s hospital, government medical centre
- ❍Private practice
- ❍Other, please specify ……..

In which country do you practice? ……

4.3 ARTÍCULO 3 - Recomendaciones para desarrollar la transición en adolescentes con enfermedades reumáticas.

TÍTULO. Estándares y recomendaciones para la atención durante la transición de jóvenes con enfermedades reumáticas de inicio en la infancia / EULAR/PReS standards and recommendations for the transitional care of young people with juvenile-onset rheumatic diseases (*Ann Rheum Dis*, 2017; 76: 639-646).

OBJETIVO. Desarrollar estándares y recomendaciones para la atención durante la transición para jóvenes con ERyME.

RESUMEN DE LOS RESULTADOS. Una vez concluida la revisión sistemática (**artículo 1**) se procedió a la discusión de los resultados obtenidos por un panel de expertos multidisciplinar constituido por reumatólogos pediátricos y de adultos, enfermeras, psicólogos y representantes de asociaciones de pacientes procedentes de toda Europa. Se propusieron un total de 12 recomendaciones relacionadas con diferentes elementos del proceso de transición y se sugirieron indicadores de calidad para cada uno de ellos. A continuación, cada una de las recomendaciones fue formulada como enunciados y se enviaron a profesionales sanitarios de unidades de reumatología y reumatología pediátrica para valorar el grado de acuerdo para cada una de las recomendaciones. También se establecieron los estándares óptimos o mínimos y el nivel de evidencia para cada recomendación. Una estrella (*) significa que es una recomendación débil con evidencia limitada. Dos estrellas (**) significa que es una recomendación débil con alguna evidencia. Tres estrellas (***) significa que es una recomendación fuerte con alguna evidencia. Cuatro estrellas (****) significa que es una recomendación fuerte con mucha evidencia.

El consenso final derivó en las siguientes **12 recomendaciones específicas** para la llevar a cabo la transición para jóvenes con ERyME:

I. **Los jóvenes y sus familias deben tener acceso a una atención multidisciplinar coordinada y de alta calidad que aborde de forma adecuada las necesidades de forma individualizada*.**

La atención debe centrarse en más aspectos que el tratamiento médico, y debe adaptarse a las necesidades presentes y futuras de cada paciente en particular, teniendo en cuenta el bienestar psicológico y social, así como los proyectos educativos y laborales.

II. **La transición debe comenzar lo más precozmente posible, ya sea en la adolescencia temprana o poco después del diagnóstico si la enfermedad comienza en la adolescencia***.**

Para los jóvenes con ERyME de inicio en la infancia, el proceso de transición idealmente debe comenzar a la edad de 11 años, y no más allá de los 14 años. Esto les permite desarrollar habilidades de autocuidado y optimizar sus posibilidades educativas y laborales. Si se realiza el diagnóstico después de los 14 años, debe iniciarse la transición de inmediato.

III. Debe haber una comunicación directa entre todos los participantes durante la transición. Antes y después de la transferencia debe haber un contacto directo entre los equipos de reumatología de las unidades pediátricas y de adultos*.

Sería conveniente la creación de redes de trabajo de reumatólogos adultos y pediátricos que trabajen juntos para garantizar una transición temprana y proactiva. Idealmente, debería haber una reunión presencial entre los jóvenes y sus familias y los equipos médicos. Si esto no fuera posible, al menos debe haber 2 contactos telefónicos o por correo electrónico entre los equipos de reumatología pediátrica y para adultos, antes y una después de la transferencia.

IV. El proceso de transición individual y el progreso se deben documentar y planificar con el joven y su familia*.

Idealmente, debe haber un proyecto de transición escrito para cada paciente. Estos documentos sirven de apoyo para el cuidado de su enfermedad y dirigirlo a una serie de servicios que pueden ser útiles. La inclusión de cualquier información médica confidencial debe discutirse con el paciente. Como mínimo, la transición debe registrarse en el historial médico.

V. Todos los servicios de reumatología deben tener un programa de transición escrito con actualizaciones periódicas*.

Debe haber un programa por escrito que detalle cómo se trasladarán los jóvenes desde la atención pediátrica a la de adultos. Estos documentos se deben actualizar cada cinco años.

VI. Deben definirse los componentes del equipo sanitario que participa en la transición, incluyendo la designación de un coordinador*.

El equipo de transición puede ser multidisciplinar e incluir a médicos, enfermeras, fisioterapeutas, terapeutas ocupacionales y/o trabajadores sociales. Es esencial que haya una persona designada en el equipo que asuma la responsabilidad de la coordinación y evaluación del proceso.

VII. Los servicios de transición deben estar centrados en los jóvenes y ser apropiados para ellos*.

Es importante adaptar la actitud y la comunicación del personal sanitario y organizar la atención sanitaria para minimizar el impacto en la vida diaria de los jóvenes. El objetivo final es permitirles tomar el control del cuidado de su enfermedad y no depender de otros.

VIII. Debe haber un informe de transferencia*.

El contenido del informe de transferencia debe ser acordado por los equipos pediátricos y adultos. Como mínimo, debe incluir el diagnóstico y un resumen de los tratamientos recibidos y el tratamiento actual,

complicaciones presentadas y otras enfermedades asociadas. Pueden añadirse más datos como situación formativa o laboral.

IX. Los profesionales sanitarios implicados deben tener una formación adecuada en enfermedades reumáticas de la infancia y en la atención a adolescentes*.

Las personas que trabajan en los equipos de transición deben conocer las enfermedades reumáticas de la infancia, su diagnóstico y tratamiento. También deben comprender los problemas generales de salud de los adolescentes y cómo podrían afectar a su enfermedad, estar capacitados para abordar cualquier problema emocional, mental o social que puedan tener los jóvenes, y poder ayudar a los jóvenes a lograr un estilo de vida saludable.

X. Debe haber financiación para los servicios de transición*.

La atención durante la transición debe prestarse en función de las necesidades individuales, y no solo de la edad o la disponibilidad de recursos. Los profesionales de la salud y los investigadores deberían tratar de garantizar que haya financiación para ayudar a los jóvenes a pasar de la atención pediátrica a la atención de adultos.

XI. Debe haber una plataforma electrónica de acceso libre donde se puedan encontrar recursos para llevar a cabo la transición*.

Pueden encontrarse de forma gratuita información y recursos útiles acerca de la transición en los sitios web de EULAR y PReS.

XII. Es necesario aumentar el conocimiento para mejorar el pronóstico de los jóvenes con ERyME de inicio en la infancia*.

Es necesario seguir investigando para mejorar la atención de los jóvenes con ERyME. Esto incluye la investigación sobre la transición, incluyendo la evaluación del éxito del proceso y el impacto de los programas de transición sobre los jóvenes.

CONCLUSIONES. Estas recomendaciones proporcionan información relevante sobre las estrategias necesarias para alcanzar resultados óptimos en la transición de jóvenes con ERyME según la evidencia disponible y la opinión de un grupo multidisciplinar de expertos. Su implementación dependerá de factores locales como los sistemas de atención sanitaria y marcos regulatorios.

EULAR / PReS standards and recommendations for the transitional care of young people with juvenile onset rheumatic diseases

Authors: Helen Foster[1], Kirsten Minden[2,3], Daniel Clemente[4], Leticia Leon[5,6], Janet E McDonagh[7], Sylvia Kamphuis[8], Karin Berggren[9], Philomine van Pelt[10], Carine Wouters[11], Jennifer Waite-Jones[12], Rachel Tattersall[13], Ruth Wyllie[14], Simon R Stones[15], Alberto Martini[16], Tamas Constantin[17], Susanne Schalm[18], Berna Fidanci[19], Burak Erer[20], Erkan Dermikaya[21], Seza Ozen[22], Loreto Carmona[23].

Affiliation: [1]Newcastle University, Institute of Cellular Medicine (Rheumatology), Newcastle, UK. [2]Children's University hospital Charité, Campus Virchow, SPZ, Berlin, Germany. [3]Epidemiology Unit, German Rheumatism Research Centre, Berlin, Germany. [4]Paediatric Rheumatology Unit, Hospital Infantil Universitario "Niño Jesús", Madrid, Spain. [5]Hospital Cínico San Carlos, Instituto de Investigación Sanitaria del Hospital Clínico, San Carlos (IDISSC), Madrid, Madrid, Spain. [6]Health Sciences, Universidad Camilo José Cela, Madrid, Madrid, Spain. [7]University of Manchester, Centre for Musculoskeletal Research, Manchester, Manchester, UK. [8]Immunology and Infectiology, Erasmus MC—Sophia, Rotterdam, The Netherlands. [9]Karolinska Universitetssjukhuset, Stockholm, Sweden. [10]Department of Paediatrics/Paediatric Rheumatology, Erasmus MC Sophia Children's, Hospital Rotterdam, Rotterdam, The Netherlands. [11]Pediatric Immunology, University Hospital Gasthuisberg, Leuven, Belgium. [12]University of Leeds, School of Healthcare, Leeds, UK. [13]University of Sheffield, School of Health and Related Research, Sheffield, Sheffield, UK. [14]Paediatric Rheumatology, Newcastle upon Tyne Hospitals NHS Foundation Trust, Newcastle upon Tyne, Newcastle upon Tyne, UK. [15]London, UK. [16]Pediatria II, Universita degli Studi di Genova Scuola di Scienze Mediche e Farmaceutiche, Genova, Liguria, Italy. [17]Department of Pediatrics, Semmelweis Egyetem, Budapest, Hungary. [18]Transitionssprechstunde am Dr. von Haunerschen Kinderspital, Ludwig-Maximilians-Universität München, Munchen, Germany. [19]Gulhane Military Medical Academy, School of Nursing, Ankara, Turkey, Ankara, Turkey. [20]Division

of Rheumatology, Department of Internal Medicine, Istanbul University, Istanbul Faculty of Medicine, Istanbul, Turkey. [21]Gulhane Military Medical Faculty, Pediatric Rheumatology Unit, FMF Arthritis, Vasculitis and Orphan Disease Research in Pediatric Rheumatology (FAVOR), Etlik, Ankara 06018, Turkey, Ankara, Turkey. [22]Pediatric Rheumatology, Hacettepe University Medical Faculty, Ankara, Turkey. [23]Instituto de Salud Musculoesquelética, Madrid, Spain.

Corresponding author: Leticia León. Instituto de Investigación Sanitaria del Hospital Clínico San Carlos (IDISSC), Hospital Clínico San Carlos, Martín Lagos s/n, Madrid, Spain. Email: lleon.hscsc@salud.madrid.org.

ABSTRACT

Objective. To develop standards and recommendations for transitional care for young people (YP) with juvenile onset rheumatic and musculoskeletal diseases (jRMD).

Methods. The consensus process involved the following: 1) Establishing an international expert panel to include patients and representatives from multidisciplinary teams in adult and paediatric rheumatology 2) A systematic review of published models of transitional care in jRMDs, potential standards and recommendations, strategies for implementation and tools to evaluate services and outcomes; 3) Setting the framework, developing the process map and generating a first draft of standards and recommendations; 4) Further iteration of recommendations; 5) Establishing consensus recommendations with Delphi methodology; and 6) Establishing standards and quality indicators.

Results. The final consensus derived 12 specific recommendations for YP with jRMD focused on transitional care. These included: high quality, multidisciplinary care starting in early adolescence; the integral role of a transition coordinator; transition policies and protocols; efficient communications; transfer documentation; an open electronic-based platform to access resources; appropriate training for paediatric and adult health care teams; secure funding to continue treatments and services into adult rheumatology; and the need for increased evidence to inform best practice.

Conclusions. These consensus-based recommendations inform strategies to reach optimal outcomes in transitional care for YP with jRMD based on available evidence and expert opinion. They need to be implemented in the context of individual countries, health care systems and regulatory frameworks.

INTRODUCTION

Transitional care, as defined by the Society for Adolescent Medicine, is "the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to adult-oriented health care systems".[1] Transition does not only focus on the administrative event of transfer of care between paediatric and adult providers; transition encompasses the process by which young people (YP) acquire skills and access resources to ensure that their physical, psychosocial, educational and vocational needs are met during transition to adulthood. [2] Adolescence and young adulthood reflect an important and unique developmental period for all YP who need education, support, guidance and planning to prepare them to be appropriately responsible and accountable for their own health and well-being as adults. [3] The same principle applies to YP with chronic illnesses (including juvenile-onset rheumatic and musculoskeletal diseases [jRMDs]) who need to acquire additional skills to independently manage their chronic illness. The case of need for transition is well described and transitional care aims to provide support and guidance so that YP can acquire the necessary skills and knowledge required to be independent, empowered and responsible adults. [4-7]

The course of jRMDs often continues into adulthood; according to population-based inception cohort studies approximately half of YP with jRMDs enter adulthood with active disease, or develop flares of disease as adults. Many YP require ongoing and often long term treatment with complex immunosuppressive regimes. [8-10] Disease related sequelae are still observed, although with modern approaches to management, many YP are transferring to adult care in clinical remission albeit on medication. [11-13] All YP with jRMDs are, in principle, at significant risk of disability, early morbidity and limitations in participation later in life. [14,15] These YP need continuous and developmentally appropriate care during and beyond adolescence to ensure optimal functioning in adulthood. However, the literature informs us that currently up to half of the YP do not make a successful transfer to adult rheumatology and are therefore at particular risk of unfavourable outcomes. [16-18]

The importance of transitional care in YP with jRMD has been increasingly acknowledged. [19,20] There is evidence regarding 'best practice' for transitional care, with emphasis on a holistic 'life course' approach to care. [21-23] Several healthcare institutions, specialties, and disease specific subgroups have developed and implemented transition programmes. [24-34] However, there is lack of clarity regarding the impact of transition programme on outcomes, and indeed, variance in what the outcomes should be. [35-40] A first consensus-based proposal regarding outcome indicators for successful health care transition was recently made by an international group of interdisciplinary health care professionals, patients and their families. [41] With rheumatology, there are significant gaps in current delivery of transition services and these include the unmet training needs for health care professionals in adolescent health and transitional care (resulting in lack of understanding and appreciation of the needs of YP), lack of transition readiness of YP (and/or of their parents/carers), and lack of robust quality indicators or cost-effective strategies. [2,36,42-48]

Despite the limitations of the existing programmes, the gaps in knowledge and the paucity of resources, there is nonetheless commitment within the rheumatology communities (both adult and paediatric), to improve existing transitional care services. There is a strong desire for rheumatology-specific guidelines for transition [48] and for these to be implemented across paediatric, adolescent and adult rheumatology health care settings.

Objectives, scope, users and overarching principles

The objective of the present initiative was to develop recommendations and standards for transitional care for YP with jRMDs, spanning ages from early adolescence (defined as 10 to 13 years), mid adolescence (14-16 years), late adolescence (17 to 19 years) to young adulthood (20 to 24 years). [49,50] These recommendations and standards are to be used to guide service development, benchmark the quality of transition services and be used by patient organisations to enhance patient expectations of care. We acknowledge that their implementation into clinical practice will be challenging and likely to be facilitated by stratification into 'essential' and 'ideal' components–**essential** defined as the *minimum* standards below which care would be

deemed unacceptable and **ideal** being the standard that is regarded as excellent "optimal" care.

The purpose of these recommendations and standards is to increase the profile of transition, optimise delivery of transitional care and improve patient experience within rheumatology across European countries. Specifically, the objectives are:

- to ensure youth friendly and developmentally appropriate care
- to improve physical, psychological, social, vocational and illness-related outcomes of YP with jRMDs
- to facilitate continuity of care within adult rheumatology
- to promote evidence-based practice in transitional care
- to facilitate clinical networks of health care professionals (paediatric and adult) who are engaged, interested and trained in the care of YP

The scope of these recommendations and standards refers to all persons involved in the care of YP with jRMDs that continue into adulthood including, but not restricted to, those in Box 1.

Although these recommendations and standards are related to the specific needs of YP with jRMDs, our expert panel endorse the American Academy of Pediatrics, the American Academy of Family Physicians, and the American College of Physicians-American Society of Internal Medicine consensus statement on health care transitions for YP with special health care needs, [51,52] position papers of the Society for Adolescent Medicine, [1, 53-55] the Canadian Paediatric Society, [56] the Royal Australasian College of Physicians, [57] and the WHO definition of adolescent-friendly health services. [58,59] In addition, we emphasise key components integral to these recommendations and standards, namely the importance of a YP focus, multidisciplinary approach with equity of access, quality of care and flexibility; the latter acknowledging both the heterogeneity of YP development and potential impact of chronic illness.

METHODS

The consensus process underwent the following stages: 1) Establishing an international expert panel to include patients and representatives from multidisciplinary teams in adult and paediatric rheumatology; 2) A systematic literature review; 3) Setting the framework, developing the process map and generating a first draft of standards and recommendations; 4) Further iteration of recommendations; 5) Developing consensus recommendations with Delphi methodology; and 6) Establishing standards and quality indicators, as suggested by the EULAR Standard Operating Procedures. [60]

The project convenors (HF, KM, LC) liaised to appoint clinical fellows (DC, LL) to work on the project. They then convened an expert multidisciplinary panel from adult and paediatric rheumatology across Europe (doctors and allied health professionals with interest in transitional care) and patient representatives (YP with jRMDs invited from existing patient groups). The aim was for the panel to reflect the diversity of Europe (namely geography, health care systems and cultures).

A systematic literature review of existing models of transitional care in jRMD was performed, with emphasis on potential recommendations, standards, strategies for implementation and tools to evaluate services and outcomes. [61]

The first 'face-to-face' meeting of the expert panel discussed results of the systematic review and agreed the following:

i) purpose of the project, timelines, roles and planned outputs

ii) the 'process map' of transitional care using MindManager™ software

iii) a draft proposal of recommendations and standards relating to different elements of the process map; the proposed list was circulated after the meeting by email to the expert group and further feedback requested.

A second 'face-to-face' meeting further refined the recommendations and standards based on feedback from the group. Appropriate quality indicators were also suggested for each recommendation and standard.

The recommendations and standards were then listed as statements. A wider audience of a total of 195 adult and paediatric rheumatology clinicians (doctors and allied health professionals) were then invited to take part in an e-survey and give opinion of their level of agreement with each statement. The e-survey was disseminated through email lists held by professional groups (such as PReS, EULAR and rheumatology societies in different countries). All responses were anonymised. Participants were asked for their level of agreement with each statement (using a 10-point Likert scale, with 0 = No agreement through to 10= Total agreement) and a ranking exercise to identify "minimal" and "optimal" standards for each recommendation. The target number of respondents for the e-survey was 100 and the level of agreement set at 80% for acceptance (lower levels were to be then discussed by the expert panel, with further iterations of the statements proposed and then a second e-survey, if needed, to be disseminated). Once agreement was reached, the methodologist (LC), together with the clinical fellows (DC, LL), graded the level of evidence for each recommendation based on the Oxford Levels of Evidence, 2011 (available at http://www.cebm.net/index.aspx?o=5653) and assigned relevant quality indicators where appropriate.

RESULTS

The recommendations and standards

The recommendations and standards, reported as "minimal/essential" and "ideal/optimal" levels of care and quality indicators are listed below; they are also presented in table format with the level of evidence and agreement reached (Table 1).

1. **YP with jRMDs should have access to high quality, coordinated transitional care, delivered through partnership with health care professionals, YP and their families, to address their needs on an individual basis.**

High quality care means holistic care (which covers medical, psychosocial, educational and vocational aspects; see box 2) with a multidisciplinary approach and is

based upon regular assessments of the disease status in the context of developmental stage, life events and personal aspirations of YP. This care needs to be 'future-focussed', albeit not limited to young adulthood, in order to ensure optimal well-being. [19,23]

Ideal- Provision of a broad set of health care services led by providers who have specialist knowledge about jRMDs and adolescent health; Essential: care providers who refer to other agencies and services who can appropriately assist with transition issues.

2. **The transition process should start as early as possible; in early adolescence or directly after the diagnosis in adolescent-onset disease.**

Specifically for childhood onset diseases, the transition process should start by early adolescence (11 years) (Ideal) or 14 years at the latest (Essential) in order to allow the development of the necessary self-care skills and optimise educational and vocational outcomes. [62,63] For YP who are diagnosed over the age of 14 years the transition process should start at the time of diagnosis with the skills and support for transition built up over time. The transition process or joint care programs should enclose early adulthood, because young adults continue to have difficulties making effective linkages with adult care. Brain development continues and risk behaviours remain and may increase in the third decade, which have to be considered by proactive and preventive care. [64]

3. **There must be 'direct' communication between the key participants (and as a minimum, to include the YP, parent/carer, and a member of each of the paediatric and adult rheumatologist teams) during the process of transition. Before and after the actual transfer there should be 'direct' contacts between paediatric and adult rheumatologist teams.**

A network of adult rheumatologists interested, engaged with and trained in adolescent rheumatology, must be identified and known to the paediatric rheumatology team. The network should work within agreed pathways to facilitate transition and expedite early, active planning of transition. Ideally, there should be a combined meeting between the young person and his or her family, the paediatric and adult health care provider. [65-67] As a minimum, there should be at least two 'direct' contacts by

telephone or email between the paediatric and adult rheumatology team (and documented in a written communication); one before and one after the transfer. Copies of written communications are to be made available to YP and families. Online supplementary table contains suggested guidelines regarding the content and format of communications at different stages of transitional care.

4. **Individual transition processes and progress should be carefully documented in the medical records and planned with YP and their families.**

Documentation should support the YP engagement and self-management skills, resilience and readiness for transfer. [68-72] This documentation should be tailored to local services, shared with the YP, and contribute to the medical summary. The inclusion of sensitive or confidential information (e.g. abortion, mental health problems) should be discussed with the YP. Ideally, there is a specific written individual transition plan, [26] which can be derived from a transition plan or passport example, such as http://www.uhs.nhs.uk/Media/Controlleddocuments/Patientinformation/Childhealth/Ready-Steady-Go-Transition-plan.pdf. [21] As a minimum, the existence of a transitional care process has to be documented in the medical records. Additional resources are listed in Supplementary material.

5. **Every rheumatology service and clinical network—paediatric and adult—must have a written, agreed, and regularly updated transition policy.**

Policies and protocols should be agreed with all major stakeholders, including YP, families and all health care professionals and as equal partners; [27,33,55] it is important to stress the need to include all specialists (and not just in rheumatology) and primary care physician(s) who are involved in the clinical care of YP with jRMD [28] Hospital or institutional managers will have to agree to these policies to facilitate appropriate resources to support their implementation within the clinical departments. As a minimum, there must be a transition policy and the documents should be updated at least every five years. It is acknowledged that there is need for flexibility of the arrangements in transition policy and care pathways at a network level.

6. **There should be clear written description of the multidisciplinary team (MDT) involved in transitional care, locally and in the clinical network. The MDT should include a designated transition coordinator.**

The team for transitional care should reflect the multidisciplinary approach, i.e. doctors and other health professionals, such as nurses, physical therapists, psychologists, occupational therapists (OT), and youth or social workers. [28,73-74] It is recognised that certain roles within transition are likely to be addressed by different members of the MDT. [75] In addition, it is acknowledged that the composition of MDTs is variable and that some members may have more than one role. Ideally, there is personal continuity in the health professionals within the MDT providing care. [43] This provision of roles and services may be shared with other specialist services and not devoted solely to jRMDs. There should be a nominated and identified member of the MDT who is responsible as transition coordinator (Essential). This person can be a nurse or other health professional and should liaise between adult and paediatric teams to ensure the coordination of care, facilitation of communication and implementation of the transitional care plan including transfer. [61]

7. **Transition services must be YP focused, be developmentally appropriate and address the complexity of YP development.**

Components of YP focused care need to include: accessibility to specialised health care, staff attitudes, communication, medical competency, guideline-driven care, age appropriate environments, and youth involvement in health care. [76] Ideally, there should be a care facility that is truly adolescent-friendly staffed by professionals with expertise in adolescent care. The care should be organised to minimise the frequency of appointments and interruption to the daily life of YP. The aim of the consultations with the MDT is to enable YP to take lead role instead of the parent / carer(s), whilst also supporting the parent / carer(s) in their changing roles. [43,77,78]

YP with jRMDs should have access to peer discussion and support through advisory group(s) and charity networks. Signposting to such groups and networks is the responsibility of the clinic where the YP attends. As a minimum, transitional care services

should be led by staff with expertise and training in adolescent and young adult healthcare.

8. There must be a transfer document.

The format and content of transfer documents should be agreed by paediatric and adult teams and with patient input where possible. The transfer document should include, as a minimum, a medical summary with the diagnosis, any comorbidities, vaccinations, any complications of disease or treatments, the professionals involved in care, current and previous treatments (with reasons for changing treatments and any adverse events). Ideally, it should also include: i) Psychosocial aspects and educational / vocational status at the time of transfer, and ii) A report on self-management skills, to include readiness for transfer and procedural pain management strategies (e.g. for joint injections with or without general anaesthesia). [79]

This transfer document may also include contributions from members of the MDT as appropriate and if relevant. Any confidential information or sensitive information to be included in the transfer document should be discussed with the YP; if needed, this should be included in a separate letter to the adult rheumatologist and be written ahead of the first consultation with adult rheumatology. Copies of the transfer document should be available to YP themselves in an easy read format if appropriate, and to all health care providers, including primary care, involved in the young person's care. [80]

9. Health care teams involved in transition and adolescent-young adult care must have appropriate training in generic adolescent health and childhood onset RMD.

All members of the clinical MDT (from adult and paediatric rheumatology) engaged in transition are to have training on adolescent health and the process of transition. [46,48] As a minimum, the key training components to be covered are: 1) jRMDs (presentation in childhood, knowledge and approaches to management), 2) Adolescence health and the impact on jRMDs, 3) Skills and knowledge to address emotional, mental health and social issues, 4) Promotion of healthy lifestyle and generic

health issues, 5) Promotion of self-management and shared decision making, 6) Communication skills with young people and their parent/carer(s).

These skills and knowledge can be acquired through different ways of learning (e.g. to include clinical experience, e-learning, and practical workshops) and should be a component of continuous professional development. There are many courses and e-learning opportunities available, such as the EULAR/PReS On-line Course in Paediatric Rheumatology (www.eular.org/edu_online_course_paediatric.cfm), the EuTEACH-European Training in effective adolescent health and care (www.unil.ch/euteach/en/home.html) or the UK Adolescent Health Project (www.e-lfh.org.uk). Reciprocal periods of training for members of the adult and paediatric rheumatology teams are advocated (Ideal).

10. There must be secure funding for dedicated resources to provide uninterrupted clinical care and transition services for YP entering adult care.

The following are regarded as essential for transitional care programmes;

i) The funding and supporting resources for care should be dependent on clinical need and should not be interrupted on sole grounds on the age of the patient. Conversely, the presence or absence of resources should not define the timing of transfer.

ii) Funding of (biological and other) therapies should continue if clinically indicated irrespective of patient age and transfer to adult care.

iii) Funding of the paediatric and adult MDTs involved in the transitional care is needed.

iv) The transition coordinator role and administration support for clinical networks must be funded.

v) The training for the MDT along with continuous professional development support must be funded

vi) The importance of adequate administrative support is likely to be a determinant of successful transfer. [45,48,80]

11. There must be a freely accessible electronic-based platform to host the recommendations, standards, and resources for transitional care.

An open resource e-platform (Essential), such as the EULAR / PReS website, to host resources to support the transition process, staff training and patient resources, would facilitate setting up of new and further development of existing transition services. All stakeholders must have access to these resources, including YP with jRMDs, their families and healthcare professionals in hospital, primary and community care involved. The resources must be endorsed (Essential) by professional bodies such as EULAR / PReS, consumer groups and charities in the respective countries.

12. Increased evidence-based knowledge and practice is needed to improve outcomes for YP with childhood onset RMD.

The expert group recognised the need for a greater evidence base to inform best practice, the best metrics for measuring "success" and "outcome" of transitional care programmes and the impact of such programmes on YP with jRMDs. [7,37,38] The agenda for research needs to include,

i) The transition programme evaluation as a complex intervention [74]

ii) The effectiveness of the transition process [61] and how this can be measured

iii) The timeliness of interventions and validation of readiness tools, [68-72,81]

iv) The outcome measures of transition [37-39,41,82] and

v) Predictors for transition outcomes

vi) Cohort studies and registries from paediatric rheumatology to extend into adult life

vii) An agreed 'core transition dataset' for routine practice in paediatric and adult rheumatology centres to inform and foster future research initiatives, facilitate a standardised approach in transitional care and enable comparative assessment of care across Europe.

Standards and quality indicators

The expert group concluded that there is a limited evidence base for outcomes of transition and for YP rheumatology services. [61] The expert group proposed, before the publication of the previous studies, key quality indicators to measure service delivery and to inform the research agenda. Supplementary table shows a list of standards and the agreed quality indicators.

In 2015, Suris *et al*. coordinated an international consensus on key elements and one indicator of a good transition. [79] In most instances, such consensus underscored items related to coordination and communication as basis for good partnership between paediatric and adult providers. Another initiative, developed during our project, obtained a clinical practice-benchmark tool for transition to adult care in the United Kingdom through a process of mapping. [83] There is no single outcome of 'successful' transition and potential indicators need to include clinical parameters (disease activity and status), patient (and family) experience (of care), psychosocial, educational and vocational status, quality of life measures, participation in adult life, engagement and attendance in adult health care, adherence to treatment, achievement of young adult developmental tasks. A recent taskforce identified, by Delphi methodology, outcomes of importance. [41] These included individual outcomes (quality of life, understanding the characteristics of conditions and complications, knowledge of medication, self-management, adherence to medication, and understanding health insurance), health services outcomes (attending medical appointments, having a medical home, and avoidance of unnecessary hospitalization), and a social outcome (having a social network). Measures need to be valid, reproducible and relevant.

DISCUSSION

This PReS / EULAR taskforce has developed the first international set of recommendations and standards for transitional care of YP with jRMDs; the aim being to facilitate high quality models of care for new and existing services, inform strategies for evaluation and define a research agenda. These were produced before the emergence of the 2016 NICE guidance on Transition in the UK (www.nice.org.uk/guidance/ng43) but reassuringly, are consistent NICE recommendations. Our methodology permitted critical appraisal of published models of care and incorporated opinion from a diverse expert specialists group including YP. [61] Our recommendations and standards set out the 'essential/minimal' and 'ideal/optimal' components of transitional care and we anticipate that such stratification will be helpful to benchmark services and facilitate implementation and evaluation.

The recommendations focus on transitional rheumatology care; however, they comprise components of high-quality transitional care derived from policy documents, guidelines relating to transitional care and the adolescent health literature. This underlines that, overall, most key elements of transitional care are generic. That is also reflected in the key elements for successful transition, which were published by Suris after the recommendations given here were agreed on. [79] Six elements were regarded as being essential by more than 70% of an international panel, of which two relate to establishing a good partnership between paediatric and adult professionals and the shared responsibility of transitional care. All six essential elements are included in the recommendations given here, which highlight the need for teams to work effectively together and engagement of different care providers within clinical networks. Addressing the challenges of 'joined up' working across paediatric and adult rheumatology and within clinical networks has also been highlighted by others. [84] Our recommendations, in agreement with other recent taskforces, [79,83] emphasise the importance of identifying key individuals, the integral role of YP and families, written communication, agreed policy, training and clarity of roles within teams. Therewith, they focus on process areas that are most in need of improvement according to care providers and consumers, such as co-ordination, guidelines, protocols, and

communication. [65] It is apparent that a transitional care pathway for YP with jRMDs can be implemented with a motivated health care team, the reorganisation of their existing work practice and available resources. [29,31] The transitional care MDT also needs specific training in adolescent medicine and adequate capacity to enable the transition care coordinator role to function. [46,48] Transition is resource consuming. The expert panel stresses the fact that without sufficient funding or reimbursement of the specific interventions, transitional care services cannot become a normal part of health care for young people. Funding is needed for specific service provision and to ensure continuity of clinical care and access to medicines after transfer to adult care based on clinical need rather than age of the patient of the provider. There are promising examples in the U.K. or Germany, where the provision of transitional care services as part of clinical practice has been funded by the government or statutory health insurance companies within defined programmes. [32] The funding for continuity of clinical care and access to medicines after transfer to adult care has to be addressed and based on clinical need rather than age of the patient of the provider. Given the importance of transition for the many YP who transfer to adult rheumatology, we strongly suggest that transitional care is included in all PReS and EULAR activities to raise awareness, promote access to training and improve skills and knowledge amongst all adult rheumatology teams.

Quality indicators and outcomes of transitional care are proposed. These transitional care outcomes are similar to most of 10 prioritized outcomes identified by a task force from the Health Care Transition Research Consortium [41] and may allow researchers to conduct focused evaluations of current processes and more detailed evaluations of interventions.

We acknowledge differences between countries in how transition may be organized within different health care systems. [65,85-87] Our recommendations are intended to be useful, widely applicable and promote transitional care. Although transitional care has received much attention in the child health community, little government attention has been paid to this complex health system issue. An analysis of policy profiles of paediatric-to-adult care transitions in six European countries revealed

that four had currently no transition policies or strategies. [87] The overarching principles seek to promote transferability to different contexts, be compliant with national regulatory guidance and facilitate local teams to work together with responsibility and accountability for services to be suitable for local needs. Our approach is similar to that proposed by others; the Spanish consensus for transition management in patients with jRMDs [88] reported a framework with similar recommendations albeit with more practical details suited to the Spanish health care system.

Transition is a time-variable process that prepares YP with jRMDs to take responsibilities for their lives and also their health issues. This process is critical in order to facilitate the actual transfer to adult care. Transition is therefore a complex process with many variables involved and the panel required considerable dialogue and indeed compromise to agree the aims, framework and process map to address such complexity. However, as the project evolved and moved forward, harmonisation became apparent and ultimately one round of Delphi was adequate to achieve high agreement.

It is clear that there are many unanswered questions in transitional care. Our recommendations highlight the need for improved evidence base to inform models of care, identify relevant outcome measures and the cost effectiveness of transitional care programmes as a complex intervention. Much work is yet to be done, but it is important to identify and ultimately deliver 'best' care for YP with jRMDs and their families, to facilitate optimal physical, psychosocial and quality of life outcomes within adulthood.

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Box 1. Individuals to whom these recommendations may prove useful.

Adult and paediatric rheumatologists
Other health care professionals: - nurses, - physiotherapists, - occupational therapists, - social workers, - psychologists, - youth workers, - primary care and other specialists (e.g., ophthalmology, dermatology, nephrology, orthopaedics) involved in share care and clinical networks
General practitioners and health care professionals working in primary care
Young people with RMD and other multisystem disease, their families and peers
Professional groups or societies working with young people with RMDs
Academics involved in Adolescent and Young Adult health research
In addition, these standards and recommendations are intended to be useful to the remit of: - Educational and vocational services - Employers and careers advisory services - Charities and support organisations - Funders of clinical services - Health policy makers - Research funding bodies - Organisers of disease and/or drug registers, and cohort studies

Table 1. Recommendations, standards, and proposed quality indicators on transitional care for young persons with RMD

Recommendations	LOE	GR	MA
1. Young people (YP) with RMD should have access to high quality, coordinated transitional care, delivered through partnership with health care professionals, YP and their families, to address needs on an individual basis.	5	D	9.6
2. The transition process should start as early as possible; in early adolescence or directly after the diagnosis in adolescent-onset disease	2b	B	8.3
3. There must be 'direct' communication between the key participants (and as a minimum, to include the YP, parent/carer, and a member each of the paediatric and adult rheumatologist teams) during the process of transition. Before and after the actual transfer there should be 'direct' contacts between paediatric and adult rheumatologist teams.	5	D	9.3
4. Individual transition processes and progress should be carefully documented in the medical records and planned with YP and their families.	5	D	9.2
5. Every rheumatology service and clinical network—paediatric and adult—must have a written, agreed, and regularly updated transition policy.	5	D	8.9
6. There should be clear written description of the multidisciplinary team (MDT) involved in transitional care, locally and in the clinical network. The MDT should include a designated transition coordinator.	5	D	8.7
7. Transition services must be YP focused, be developmentally appropriate and address the complexity of YP development.	5	D	9.4
8. There must be a transfer document.	5	D	9.4
9. Health care teams involved in transition and adolescent-young adult care must have appropriate training in generic adolescent care and childhood onset RMD.	5	D	9.5
10. There must be secure funding for dedicated resources to provide uninterrupted clinical care and transition services for YP entering adult care.	5	D	9.4
11. There must be a freely accessible electronic-based platform to host the recommendations, standards, and resources for transitional care.	5	D	9.4
12. Increased evidence-based knowledge and practice is needed to improve outcomes for YP with childhood onset RMD.	5	D	8.5

Abbreviations: LOE, level of evidence; GR, grade of recommendation; MA, mean agreement (0-10)

Box 2. Aspects considered as part of holistic care.

Medical aspects

- identification of medical needs, addressing any issues
- ensuring continuity of provision of high quality care
- providing generic and disease-specific information
- health promotion, anticipatory guidance
- health behaviour (e.g. health literacy, experimentation and risk behaviour), negotiating most appropriate ways to ensure adherence to treatment
- knowledge and skills in areas listed above

Psychosocial aspects

- identifying individual needs, risk and protective factors (e.g. HEADSS),(49, 50)
- providing support or referring young people to specific agencies
- ensuring a social life that is equivalent to those of peers
- ensuring support to cope with disease/treatment
- providing advice and/or additional sources of support
- promoting skills in assertiveness, resilience, self-care, self-determination, and self-advocacy

Educational and vocational aspects:

- addressing future career prospects
- developing skills in disclosure
- support in preparing for work readiness,
- informing about where to get information (recommend: career advisors, appropriate agencies, charity websites)
- addressing work experience and encouraging young people to gain relevant experience
- offer appropriate information, support and advice (support groups, volunteer services)
- liaisons with educational institutions
- informing about rights and obligations, benefits and opportunities to adapt working (place, time)

(Supplementary material)

Selected resources that may aid designing and maintaining transitional services.

Web site	Description	Useful to
TRAINING RESOURCES		
http://www.unil.ch/euteach/home.html	Provides objectives, strategies and resources for trainers and teachers.	Healthcare professionals
http://www.e-lfh.org.uk/programmes/adolescent-health/	The Adolescent Health Programme (AHP) is an e-learning programme for all healthcare professionals working with young people.	Healthcare professionals
http://www.talklab.nhs.uk	TalkLab's Better Conversations programme aims to improve the sometimes complex, three-way consultations doctors have with young people and their parents	Health care professionals, young people and families
http://www.usc.edu/adolhealth/	This site was developed for use by health care professionals involved in either the teaching of adolescent health or clinical care of adolescents and young adults. The material includes cases, questions, weblinks and references.	Healthcare professionals
GUIDANCE		
http://www.cps.ca/en/documents/authors-auteurs/adolescent-health-committee#practice-points	This website provides health professionals information needed to make informed decisions about the well being of children and youth. Parents, journalists and other stakeholders may also find this site useful.	Healthcare professionals, young people and families.
http://www.adolescenthealth.org/Clinical-Care Resources/Clinical-Care-Resource-Guides.aspx	This site provides resources to adolescent and young adult health care providers and youth serving professionals. Include clinical care guidelines and resources. Most of the resources included are applicable in the U.S. and some are internationally applicable.	Healthcare professionals, young people

Web site	Description	Useful to
https://www.nice.org.uk/guidance/ng43	This guideline of the National Institute for health and Care Excellence includes recommendations for transition.	Healthcare professionals, young people and families.
PROFESSIONAL ORGANISATIONS		
http://www.yphsig.org.uk/	Home page of the Young People's Health Special Interest Group of the Royal College of Paediatrics and Child Health, UK	Healthcare professionals
http://www.adolescenthealth.org	Home page of the Society for Adolescent Health and Medicine	Healthcare professionals
http://www.iaah.org	Home page of the International Association for Adolescent Health	Healthcare professionals
TRANSITIONAL CARE RESOURCES		
http://www.gottransition.org/	Include a complete compilation. Serve as a clearinghouse for current transition information, tools, and resources. Also in Spanish.	Healthcare professionals, young people and families.
http://depts.washington.edu/healthtr	Include transition resources such as websites, checklists and tools for teens and young adults with disability or chronic illness.	Healthcare professionals, young people and families.
http://chfs.ky.gov/ccshcn/ccshcntransition.htm	Interesting website with Transition Resources, ages adapted	Young people and families
http://www.chla.org/my-voice-adolescent-transition-program	Include information about Adolescent Transition Program	Young People and families
http://www.floridahats.org/	A health care transition tool box contains documents and links to a variety of local, state and national resources. Materials for youth, families and professionals are organized in several categories.	Healthcare professionals, young people and families.

Web site	Description	Useful to
http://www.sickkids.ca/good2go/	This site provides resources to youth, families, and health care professionals.	Healthcare professionals, young people and families.
http://www.ontracbc.ca/	Includes information about transition: preparation, transfer, and attachment to adult services. Includes toolkits for youth, parents/families, and care providers	Patients, families and health care professionals
http://www.transitioninfonetwork.org.uk	Provides information about the transition process that is useful to disabled young people, parents/carers and professionals.	Healthcare professionals, patients and families.
http://www.chimat.org.uk/transitions	Resources and evidence relating to young people's transition process into the adult world. It is regularly updated	Healthcare professionals, patients and families.
http://www.uhs.nhs.uk/OurServices/Childhealth/TransitiontoadultcareReadySteadyGo/Transitiontoadultcare.aspx	Description of the Ready Steady Go transition programme. Include resources for health professionals and useful links	Young people, families and health care professionals
http://www.rch.org.au/transition/	Transition Support Service of Royal Children's Hospital Melbourne, Australia. Includes fact sheets and tools, links, publications and others.	Healthcare professionals, young people and families.
RHEUMATOLOGY-SPECIFIC		
http://www.bspar.org.uk/adolescent-rheumatology	Excellent website of British Society for Paediatric and Adolescent Rheumatology Include a complete compilation of useful resources for young people and professionals.	Healthcare professionals, young people and families
Paediatric Musculoskeletal Matters http://www.pmmonline.org	Free website explaining the essentials of rheumatic diseases in children and young people and useful for support and training of shared care providers.	Health care professionals

Web site	Description	Useful to
http://www.healthtalk.org/young-peoples-experiences/arthritis/transition-moving-adolescent-or-adult-services	This website provides free, reliable information about health issues, by sharing people's real-life experiences.	Young people
http://www.jatransition.org/	Website designed to prepare youth with rheumatic diseases and their families for the transition to healthy adulthood.	Young people and families
http://www.arthritisresearchuk.org/youngpeople	Includes practice information for young patients, parents and also for patient school teachers.	Young people and families
http://www.jia.org.uk/transition	Include useful resources about moving from paediatric to adult care. Includes a specific guide for parents	Young people and families
http://www.kidsgetarthritistoo.org/living-with-ja/daily-life/transition/	Specifics resources for parents.	Families
http://www.hopkinsarthritis.org/patient-corner/transitioning-the-jra-patient-to-an-adult-rheumatologist/	Explanation about Transitioning the JRA Patient to an Adult Rheumatologist: patient view	Young people
http://www.jong-en-reuma.nl/	Website created for and by young people with rheumatic diseases. In Dutch	Young people
http://www.reuma-uitgedaagd.nl/	Information for young people with rheumatic diseases. In Dutch	Young people

Standards at national level

1. Adolescent and young adult care (including transition) is included in national training syllabuses (adult + paediatric) for all allied health professionals and doctors in adult and paediatric rheumatology.
2. There is adequate IT infrastructure to facilitate communication between paediatric and adult rheumatology teams.

Standards at unit level (minimum + ideal)

1. There is a professional with a designated role for transition (i.e. 'transition coordinator') working between paediatric and adult rheumatology services
2. There is a written transition policy agreed with paediatric and adult rheumatology networks and their supporting infrastructure (e.g. management teams, insurers, etc)
3. There is a clinical transfer summary template agreed by paediatric and adult rheumatology
4. There is adequate IT infrastructure to facilitate data collection, recording of outcome and quality indicators, and facilitating communication between paediatric and adult rheumatology teams
5. There is evidence of young people being actively involved in service design and development
6. There is a research programme on transition and adolescent and young adult care

Standards at patient level (minimum + ideal)

1. The YP attends the first two appointments in adult rheumatology
2. The YP receives a clinical transfer summary produced by paediatric rheumatology and agreed by the young person, and this is received by adult rheumatology and primary care provider for every YP
3. Outcome measures:
    a. achievement of transition readiness,
    b. disease knowledge,

c. improved medication adherence
d. disease control
e. quality of life
f. Participation
g. young person or family satisfaction
h. self-advocacy
i. self-management readiness
j. resilience / independent living
k. work stability and employment

Quality indicators

To test in the setting before and after the implementation of the health service.

1. % Lost to follow-up after 1 year of transfer
2. % no shows on visits in adult rheumatology after 1 year of transfer
3. YP and family satisfaction with transfer process (e.g. Mind the Gap scale)(51-53)
4. Satisfaction of members of the MDT
5. The proportion of YP with a complete transfer document
6. The number of contacts documented in the year before and after transfer for each YP
7. An agreed written policy is in place
8. The proportion of MDT staff completing the transition training programme
9. An IT platform is available to host resources, tools, guidelines and policy documents
10. Outcome measures:
    a. achievement of transition readiness
    b. disease knowledge
    c. improved medication adherence
    d. disease control
    e. quality of life

f. Participation

g. young person or family satisfaction

h. self-advocacy

i. self-management readiness

j. resilience

k. independent living

l. work stability and employment

Suggested content and format of communication at different levels of transitional care.

1) Communication between health professionals and patient: a) Developmentally-appropriate language, YP-oriented i) honest, realistic and jargon free ii) supportive dialogue iii) assisting young people to develop the communication skills that are necessary to manage and navigate services effectively iv) establishing a positive therapeutic relationship that enables young people to express opinions and make informed decisions about care b) Offering opportunities (enough time, independent of caregivers/parents, appropriate environment, talk to peers) for communication c) Developmentally-appropriate format (written, verbal, phone calls, emails, texting, referring to other forms of communication), d) Explain and assure confidentiality (e.g. before discussing sensitive topics)
2) Communication between health professionals and family: It must be in a format that can be understood, and family-oriented: i) appropriate verbal and non-verbal communication ii) referring to other forms of communication iii) acknowledging parents' knowledge base and pivotal role in caring iv) offering available supporting services v) respect rights to confidentiality of the young person

3) Communication between health professionals:

The members of the adult and paediatric rheumatology teams will have direct contact as a minimum by telephone or email before the handover of the young person, and document it. There should be at least two direct contacts between the paediatric and adult rheumatologist during the process of transition:

i) one member of the paediatric team: announcing transfer of patient
ii) one member of the adult team: feedback confirming that the patient has been seen for at least the first two visits in adult care.

5 DISCUSIÓN GENERAL

Nuestra revisión sistemática **(artículo 1)** confirmó que hay relativamente pocos modelos bien descritos para la transición de adolescentes con enfermedades reumáticas desde las unidades de reumatología pediátrica hasta su asistencia en las unidades de reumatología de adultos, y, en general, pocos datos sobre los resultados del proceso de transición y sobre que variables influyen en que ésta se realice de una forma exitosa. El nivel de programación de la transición en reumatología pediátrica es variable y depende de muchos factores que, a menudo, están relacionados con el sistema de asistencia sanitaria o los recursos disponibles a nivel local.

Sin embargo, muchas de las recomendaciones obtenidas para la transición en enfermedades reumáticas son generales y compartidas con otras enfermedades crónicas **(artículo 3)**. Esto se debe a que los problemas que presentan los jóvenes durante la transición son comunes, con independencia de la enfermedad que padezcan. Algunas modificaciones pueden ser necesarias para adaptarlo a los sistemas sanitarios de diferentes países, en los que pueden variar el fin de la edad de atención en unidades pediátricas (14-18 años) o el sistema de financiación.

Establecer por escrito un **programa de transición** acordado por los diferentes participantes en el proceso ofrece numerosas ventajas, entre ellas, la posibilidad de evaluar los resultados e introducir las modificaciones que se consideren necesarias. Sin embargo, según los resultados de nuestra encuesta, muchos de los servicios ofrecen un proceso de transición informal **(artículo 2)** y, de hecho, pocas publicaciones sobre programas de transición en enfermedades reumáticas han descrito específicamente los componentes del programa individual, los procedimientos que se realizan y la organización del proceso de transferencia **(artículo 1)**. El programa de transición de cada centro debe ser actualizado periódicamente, al menos una vez cada 5 años, y debería incluir al menos los elementos recogidos en la Tabla 7.

Tabla 7. Contenidos esenciales de un programa de transición

- Definición de transición
- Nombre y función de los miembros de los profesionales sanitarios implicados, incluyendo el coordinador de la transición
- Edad a la que se desarrollan las actividades:
  - Primera discusión con los adolescentes e inicio del programa de transición
  - Transferencia a unidad de adultos
- Plan de transición individualizado:
  - Identificación de necesidades y expectativas de pacientes y familiares
  - Establecimiento de objetivos y metas
  - Recursos educativos disponibles y estrategias para alcanzar los objetivos
- Elementos del proceso de transferencia:
  - Contacto con unidad de reumatología de adultos antes y después de la transferencia
  - Informe de transferencia
  - Implicación con atención primaria
- Evaluación del proceso de transición y revisión periódica para identificar puntos fuertes a reforzar y áreas de mejora
  - Adquisición y/o afianzamiento de habilidades y conocimientos sobre enfermedades reumáticas y sus tratamientos
  - Calidad de vida de los pacientes
  - Satisfacción de pacientes, familiares y profesionales sanitarios
  - Continuidad de cuidados en unidad de adultos

No es infrecuente que la transición se retrase hasta uno o dos años antes de la transferencia a las unidades de adultos. Sin embargo, se ha demostrado que **iniciar la transición durante la adolescencia temprana** (11-12 años) proporciona a los pacientes unos recursos mayores en conocimientos y habilidades, que a su vez dan lugar a unos mejores resultados a largo plazo (54). Este inicio temprano de la transición da más tiempo a los jóvenes para su entrenamiento para las unidades de adultos y permite

adaptar el ritmo según las circunstancias personales y sociales de cada uno de ellos. Esta flexibilidad en el proceso tiene en cuenta la variabilidad en el desarrollo adolescente y el impacto que tienen las enfermedades crónicas en la adolescencia. En los casos en los que la enfermedad comience después de los 14 años, el proceso de transición debe comenzar lo antes posible tras el diagnóstico.

Sorprende que la recomendación que hace referencia al inicio precoz de la transición **(artículo 3)** es la que menos acuerdo alcanza de las formuladas, a pesar de ser la que se sostiene con mayor evidencia. Esto puede ser debido a una confusión entre los términos de transición, un proceso gradual de empoderamiento por parte de los adolescentes, y transferencia, un evento aislado de movimiento de los pacientes desde las unidades pediátricas a los servicios de adultos. De este modo, aunque es recomendable tener una unidad de adultos de referencia, el proceso de transición tampoco debe retrasarse en aquellos casos en los que no haya un reumatólogo adulto identificado o disponible.

El equipo sanitario puede incluir a médicos, enfermeros, fisioterapeutas, psicólogos, terapeutas ocupacionales o trabajadores sociales. Contar con un **equipo multidisciplinar** facilita el abordaje óptimo de todos los aspectos que afectan a los adolescentes, pero su disponibilidad es variable dependiendo de los recursos con los que cuenta cada centro. Se necesita al menos un médico de la unidad pediátrica y un médico de la unidad de adultos que pueden asumir la asistencia conjunta de los pacientes, mientras que el resto de los colaboradores depende de los recursos específicos de cada uno de los centros. Es fundamental que uno de los componentes del equipo tome la responsabilidad de coordinar el proceso de transición, incluyendo la implementación y evaluación del programa. Esta figura de **coordinador de la transición** es un médico o un enfermero en la mayoría de los casos **(artículos 1 y 2)**, aunque puede ser cualquiera de los profesionales sanitarios implicados que tenga la formación adecuada y trabaje en colaboración con el equipo médico.

Para implementar un **plan de transición individualizado (artículo 3)** es importante conocer las necesidades de cada uno de los adolescentes con enfermedades

reumáticas, no solo las relacionadas con el conocimiento sobre su enfermedad o los tratamientos que reciben sino considerando también nivel de autonomía, estilo de vida o bienestar emocional. Dado que los jóvenes pueden ser reticentes a comenzar conversaciones sobre estos temas, el profesional sanitario debe tomar la iniciativa para poder asesorarlos. Para ello pueden utilizarse cuestionarios a lo largo del proceso de transición (Anexos 1, 2 y 3), resolviendo posteriormente de forma específica los apartados en los que el adolescente se vea inseguro o más necesitado de ayuda o recursos (p.ej., derivar al psicólogo en caso de detectar trastornos del estado de ánimo). El progreso de cada adolescente debe quedar documentado en la historia clínica, aunque la inclusión de información sensible (p.ej., problemas sociales como maltrato escolar) debe ser comentada previamente con ellos.

Además de estos cuestionarios se debe proporcionar recursos a los adolescentes y padres en forma de folletos informativos, acceso a páginas web o, idealmente, en forma de talleres que se imparten en grupo. Estos talleres pueden facilitar el intercambio de opiniones en un ambiente distinto de las consultas médicas y están enfocados a dar información y soporte a todos los aspectos de interés en la adolescencia, siendo útil seguir el esquema HEADSS (55): relaciones familiares y vida cotidiana (*Home*), educación y orientación profesional (*Education and Employment*), actividad física (*Activity*), estilo de vida incluyendo conductas de riesgo y consumo de sustancias (*Drugs*) y emociones y relaciones de pareja (*Stress, emotions and Sexuality*). Los conocimientos y habilidades sobre los que se debe formar a los adolescentes en los programas de transición se recogen en la Tabla 8.

Tabla 8. Conocimientos y habilidades en los programas de transición.

- Conocimientos para la transición
  - Definición y propósito de la transición
  - Enfermedad reumática: diagnóstico, manejo y evolución, distinción de recaídas
  - Tratamientos empleados: nombres, dosis, efectos secundarios, justificación de los mismos, riesgo de no adherencia
  - Pruebas diagnósticas y procedimientos
  - Términos médicos habituales
  - Estilo de vida saludable: ejercicio, alimentación, exposición solar
  - Impacto de alcohol y drogas sobre el tratamiento
  - Impacto de la enfermedad en vida sexual y planificación familiar
  - Consejos sobre tatuajes o piercings
  - Soporte social y grupos de apoyo
  - Fuentes fiables de información

- Habilidades para la transición
  - Autonomía: para realizar consultas médicas sin la presencia de los padres, para concertar citas, para tomar decisiones sobre los tratamientos,…
  - Capacidad para tomar o administrarse los tratamientos prescritos
  - Adherencia a los tratamientos y a las revisiones médicas
  - Manejo de la fatiga y el dolor
  - Comunicación con los profesionales sanitarios
  - Comunicación con familiares y amigos
  - Resiliencia, manejo del estrés y emociones

Para proporcionar una asistencia integral a los adolescentes con enfermedades reumáticas durante la transición a la edad adulta lo ideal sería disponer de una unidad específica o unidad de transición. Si no es posible, se debe realizar una práctica asistencial centrada en el adolescente, adecuando los espacios de atención a los mismos y cuidando la actitud y comunicación por parte de los profesionales sanitarios **(artículo 3)**.

Es necesario **fomentar la empatía y el respeto mutuo**, evitando prejuicios a la hora de valorar a los adolescentes y todo tipo de conductas que puedan interpretarse como paternalismo. También se debe asegurar y garantizar la **confidencialidad**, ya que los adolescentes no confiarán en su médico si sospechan que la información que aportan se la transmitirán a sus padres. Para **promover la autonomía** de los pacientes y disminuir la dependencia de los padres se debe recomendar acudir a las revisiones médicas sin los padres. De esta manera se desarrollan las habilidades necesarias para ser una parte activa en su cuidado de salud y se mejora la capacidad de comunicación, la toma de decisiones y la negociación. Por lo general, los profesionales sanitarios consideran que el adolescente está preparado para ser visto de forma independiente a una edad más temprana que sus padres, por lo que debería buscarse un acuerdo con ellos e incorporar **visitas independientes** de forma gradual. Para saber el grado de implicación de los padres en el proceso de transición puede utilizarse el cuestionario recogido en el Anexo 4.

Es importante habilitar los espacios de consulta, de forma que se trate adecuadamente la intimidad (puertas o cerrojos, biombos o cortinas, sábanas para cubrirse,etc.). También puede concentrar las revisiones de los adolescentes y jóvenes en un día concreto para que se puedan relacionar en la sala de espera personas de su edad con circunstancias similares. Esto puede ser especialmente recomendable cuando los pacientes sean atendidos en las unidades de reumatología de adultos. La duración, la frecuencia y los horarios de las consultas deben adecuarse en lo posible a los adolescentes para minimizar el impacto en su vida diaria.

La clave para el buen funcionamiento de una unidad de transición se basa en la competencia profesional, las cualidades y la personalidad de los profesionales sanitarios. Por ello, todo el equipo sanitario debe tener una **formación adecuada** tanto en las enfermedades reumáticas en la infancia, como en el trato con adolescentes, sabiendo abordar de forma adecuada problemas emocionales o sociales **(artículo 3)**.

El cambio a la unidad de adultos o **transferencia** debe consensuarse entre reumatólogos de la unidad pediátrica y la unidad de adultos, que deben mantener una comunicación directa entre ellos, los pacientes y sus familiares **(artículo 3)**. Idealmente, el reumatólogo de la unidad de adulto se encuentra con el paciente y la familia antes de la transferencia, estando presente en las revisiones médicas en la unidad pediátrica o en la unidad de transición. Cuando no sea posible, al menos debe existir un contacto telefónico entre los dos equipos médicos, antes y después del cambio a la unidad de adultos. El momento del cambio puede llevarse a cabo en una fecha concreta conocida por todos con suficiente antelación (p.ej., al cumplir 16-18 años), aunque lo recomendable es que este momento sea flexible y adaptado a las circunstancias individuales de cada paciente, una vez que se hayan cumplido los objetivos de la transición y evitando hacerlo durante los periodos de actividad de la enfermedad. Para ello puede ser útil una lista de comprobación como la que se presenta en el Anexo 5 **(artículo 1)**.

Toda la **información relevante** (diagnostico, tratamientos pasados y actuales, complicaciones, problemas psicosociales, etc.), debe ser recogida **en el informe clínico** que deben recibir en la unidad de adultos **(artículo 1, 2 y 3)**. Debe asegurarse la continuidad de la asistencia sanitaria y de los tratamientos durante el movimiento entre las unidades. En algunos sistemas sanitarios en los que solo está asegurada la asistencia pediátrica puede ser necesario buscar financiación adicional para asegurar esta continuidad asistencial. Solo se considera la transferencia completa cuando los pacientes continúan de forma efectiva su seguimiento en la unidad de adultos (p.ej., acuden a más de 2 visitas consecutivas).

Finalmente, en la mayoría de los programas existe una falta de medidas objetivas del proceso y de los resultados conseguidos **(artículo 1)**, y es difícil saber que elementos individuales de una intervención compleja como son los programas de transición son los que tienen más relevancia para conseguir que la transición sea considerada satisfactoria. Es necesario saber si los programas de transición consiguen mejorar el pronóstico a largo plazo de los pacientes o si realmente existen resultados positivos a nivel emocional y social, así como cuál es el mejor método para enseñar a los

adolescentes los conocimientos y habilidades que necesitan, incluyendo medidas de coste/efectividad. Entre los indicadores que pueden incorporarse en la evaluación del proceso de transición se incluyen: parámetros clínicos (actividad de la enfermedad, capacidad funcional, daño estructural), estado psicosocial y educacional, calidad de vida, adherencia a los tratamientos, continuidad del seguimiento en la unidad de adultos y satisfacción con el proceso por pacientes, familiares y profesionales sanitarios.

6 CONCLUSIONES

1. Los programas de transición de mayor calidad contienen seis puntos críticos:
    a. un programa de transición estructurado y por escrito
    b. una planificación individualizada del proceso, con flexibilidad en su implementación y duración
    c. un comienzo precoz
    d. un coordinador que, junto con el paciente y la familia, asuma la responsabilidad de la transición y la evaluación de resultados
    e. la promoción de conocimientos y habilidades a jóvenes y sus padres mediante la educación y formación de estos
    f. la transmisión de la información relevante para el equipo de adultos y un calendario de transferencia flexible.
2. Faltan indicadores estandarizados y medidas de resultado que permitan evaluar adecuadamente el proceso de la transición en jóvenes con ERyME.
3. El número de programas de transición estructurados publicados en el campo de las ERyME es limitado en comparación con otras enfermedades crónicas.
4. La implementación de programas de transición está influenciada por los recursos disponibles a nivel local.
5. Existe un fuerte compromiso de reumatólogos pediátricos y de adultos para mejorar la provisión de cuidado de transición existente.
6. Las recomendaciones de EULAR / PReS son oportunas e importantes como un catalizador necesario para el cambio dentro de la comunidad de reumatología siendo los puntos más importantes:
    a. Los niños y adolescentes con ERyME deben ser preparados de forma adecuada para ser responsables de su salud una vez que continúen sus cuidados en las unidades de adultos.

i. Este proceso de transición debe comenzar lo antes posible, idealmente a los 11 años,

ii. y ser flexible en su desarrollo para adaptarse a las necesidades de cada paciente.

b. Es necesario elaborar un programa de transición estructurado y acordado por todos los participantes del proceso.

i. Deben quedar claros las actividades a realizar, el cronograma y las funciones de cada uno de los miembros del equipo sanitario multidisciplinar.

ii. La figura de un coordinador de la transición asegura la implementación del programa y permite la revisión, actualización y evaluación de este.

iii. Durante el proceso de transición debe establecerse una comunicación directa entre los profesionales sanitarios de unidades pediátricas y de adultos, los adolescentes y sus familias.

iv. Debe asegurarse la adquisición de los niños y adolescentes con enfermedades reumáticas de los conocimientos, habilidades y competencias necesarios para cuidar de su enfermedad por sí mismos.

v. El progreso de cada adolescente y joven debe registrarse en la historia clínica y debe formar parte del informe de trasferencia, en el que deben constar los datos fundamentales que deben recibir los reumatólogos en las unidades de adultos.

c. Los servicios en los que se desarrollan estos programas de transición deben estar centrados en el adolescente, siendo imprescindible la formación de los profesionales tanto en el manejo de los problemas habituales en esta edad y de las enfermedades reumáticas que se inician en la infancia.

d. Debe asegurarse un cuidado ininterrumpido de los jóvenes durante el traslado entre las unidades pediátricas y de adultos, garantizando los recursos necesarios para la asistencia y la administración de los tratamientos durante este periodo.

e. Por último, para mejorar la atención de niños y adolescentes con enfermedades reumáticas, es necesario medir las variables que contribuyen al éxito de los programas de transición y el impacto que estos programas tienen estos pacientes.

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8 ANEXOS.

8.1 Anexo 1. Cuestionario de evaluación de conocimientos y habilidades (adolescencia temprana).

Conocimientos y habilidades	**SI**	**Necesitaría ayuda / consejo sobre este tema**	**Comentarios**
CONOCIMIENTOS			
Puedo describir mi enfermedad			
Sé las medicinas que tomo (nombre, dosis, frecuencia, …)			
Sé cómo mi enfermedad puede afectar a mi desarrollo			
Entiendo las diferencias entre unidades pediátricas y de adultos			
Entiendo lo que significa el proceso de transición			
AUTONOMÍA			
Me siento preparad@ para ser vist@ sin mis padres en el futuro			
Hago preguntas al médico			
Puedo vestirme y asearme sin ayuda en casa			
Puedo preparar mi propia comida			
Soy capaz de pasar la noche fuera de casa			
Soy capaz de usar el trasporte público para moverme por mi entorno			
Sé a qué quiero dedicarme en el futuro cuando acabe el colegio			

Conocimientos y habilidades	**SI**	**Necesitaría ayuda / consejo sobre este tema**	**Comentarios**
ESTILO DE VIDA			
Sé que el ejercicio es bueno para mi salud en general y para mi enfermedad			
Sé que cuidar la alimentación es bueno para mi salud			
Entiendo los riesgos del alcohol y el tabaco para mi salud			
Sé cómo y dónde buscar información sobre sexualidad			
EMOCIONES			
Estoy contento			
Tengo a alguien cercano con quien hablar cuando estoy triste o enfadado			
Sé cómo responder frente a comentarios provocativos y/o malintencionados			

Por favor, añade a continuación aquello que consideres que necesitas consejo o ayuda que no haya sido mencionado anteriormente

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8.2 Anexo 2. Cuestionario de evaluación de conocimientos y habilidades (adolescencia media).

Conocimientos y habilidades	SI	Necesitaría ayuda / consejo sobre este tema	Comentarios
CONOCIMIENTOS			
Entiendo los términos médicos y los procedimientos relativos a mi enfermedad			
Sé para qué sirven las medicinas que tomo y sus efectos secundarios			
Soy responsable de la medicación que tomo y sé cuándo necesito más recetas			
Sé cuándo tengo cita			
Conozco la función de cada uno de los médicos y sanitarios que me atienden			
Conozco el hospital/centro al que iré cuando sea adult@			
Conozco los recursos que existen para adolescentes con enfermedades reumáticas			
AUTONOMÍA			
Me siento preparad@ para ser visto en la consulta sin mis padres y hacer mis propias preguntas			
Entiendo mi derecho a la confidencialidad			
Puedo vestirme y asearme sin ayuda en casa y preparar mi propia comida			
Soy capaz de saber que medicinas llevar y como trasportarlas si me voy de viaje			
Conozco la posible influencia de mi enfermedad en los estudios o trabajos que me interesan			

Conocimientos y habilidades	**SI**	**Necesitaría ayuda / consejo sobre este tema**	**Comentarios**
Soy capaz de usar el trasporte público para moverme por la ciudad			
ESTILO DE VIDA			
Hago ejercicio regularmente			
Cuido mi alimentación			
Entiendo los riesgos del alcohol, tabaco y otras drogas para mi salud			
Sé cómo y dónde buscar información sobre sexualidad			
Entiendo los riesgos de un embarazo con mi enfermedad y las medicinas que tomo			
EMOCIONES			
Estoy contento			
Puedo hablar con otras personas de mi enfermedad			
Sé cómo manejar emociones como la ira, ansiedad o tristeza			

Por favor, añade a continuación aquello que consideres que necesitas consejo o ayuda que no haya sido mencionado anteriormente

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8.3 Anexo 3. Cuestionario de evaluación de conocimientos y habilidades (adolescencia tardía).

Conocimientos y habilidades	SI	Necesitaría ayuda / consejo sobre este tema	Comentarios
CONOCIMIENTOS			
Conozco mi enfermedad y los tratamientos que se utilizan			
Controlo y administro la medicación que necesito			
Organizo mis citas			
Llamo al hospital cuando tengo dudas sobre mi enfermedad / tratamientos			
Conozco los recursos que existen para adolescentes con enfermedades reumáticas			
Sé el procedimiento para continuar mi cuidado en el servicio de adultos			
AUTONOMÍA			
Puedo entrar a la consulta solo			
Entiendo mi derecho a participar en la toma de decisiones sobre los tratamientos y procedimientos que recibo			
Soy independiente en casa			
Soy capaz de planificar y organizar lo necesario para un viaje de varios días			
Soy capaz de usar el trasporte público para moverme por mi ciudad			
He comenzado estudios/trabajo/he participado como voluntario			
ESTILO DE VIDA			
Hago ejercicio regularmente			
Cuido mi alimentación			

Conocimientos y habilidades	**SI**	**Necesitaría ayuda / consejo sobre este tema**	**Comentarios**
Entiendo los riesgos del alcohol, tabaco y otras drogas para mi salud			
Sé cómo y dónde buscar información sobre sexualidad			
Entiendo los riesgos de un embarazo con mi enfermedad y las medicinas que tomo			
EMOCIONES			
Estoy contento			
Puedo hablar con otras personas de mi enfermedad			
Sé cómo manejar emociones como la ira, ansiedad o tristeza			

Por favor, añade a continuación aquello que consideres que necesitas consejo o ayuda que no haya sido mencionado anteriormente

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8.4 Anexo 4. Cuestionario de evaluación de conocimientos y habilidades (padres).

Nombre del padre_____

Nombre del paciente_____

Fecha_____

	SI	NO	NO APLICABLE
CONOCIMIENTOS Y HABILIDADES			
Entiendo el significado y el objetivo de la transición			
Sé que médicos y profesionales sanitarios atienden a mi hij@ y su función			
Sé cómo influye la enfermedad de mi hij@ durante la adolescencia			
Conozco el pronóstico a largo plazo de la enfermedad de mi hij@			
Conozco los recursos que existen para niños y adolescentes con enfermedades reumáticas			
Entiendo las diferencias entre los servicios pediátricos y de adultos			
Conozco el hospital/centro al que acudirá mi hij@			
AUTONOMÍA			
Intento que mi hij@ participe en las revisiones médicas y en la toma de decisiones			
Enseño a mi hij@ sobre cómo ser responsable de su medicación en casa			
Fomento la independencia de mi hij@ para vestirse, asearse, alimentarse,… en casa			
ESTILO DE VIDA			
Fomento que mi hijo haga ejercicio y tenga una alimentación variada			
Entiendo los riesgos del alcohol, tabaco y otras drogas para la salud de mi hij@			
Entiendo los riesgos de un embarazo con la enfermedad de mi hij@ y las medicinas que toma			

Por favor, añade a continuación aquello que consideres que necesitas consejo o ayuda que no haya sido mencionado anteriormente

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8.5 Anexo 5. Lista de comprobación/verificación en la transferencia.

LISTA DE COMPROBACIÓN/VERIFICACIÓN	SI	NO
Enfermedad controlada y estable		
Conocimiento de la enfermedad y de los tratamientos que recibe		
Capacidad para tomar decisiones acerca de su enfermedad, organizar sus citas y las medicaciones que necesita		
Se ha establecido contacto con una Unidad de Reumatología de adultos		
Se ha elaborado un informe de transferencia		
Se ha asegurado la continuidad del tratamiento durante el proceso		
Se han realizado 2 visitas en la unidad de Reumatología de adultos		

8.6 Anexo 6. Artículos

Clemente et al. Pediatric Rheumatology (2017) 15:49
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Pediatric Rheumatology

SHORT REPORT Open Access

Transitional care for rheumatic conditions in Europe: current clinical practice and available resources

Daniel Clemente[1], Leticia Leon[2], Helen Foster[3], Loreto Carmona[4] and Kirsten Minden[5*]

Abstract

Objective: To assess European pediatric rheumatology providers' current clinical practices and resources used in the transition from child-centered to adult-oriented care.

Methods: European pediatric rheumatologists were invited to complete a 17-item anonymized e-survey assessing current transition practices, transition policy awareness, and needs in advance of the publication of EULAR/PReS recommendations on transition.

Results: The response rate was 121/276 (44%), including responses from 115 centers in 22 European Union countries. Although 32/121 (26%) responded that their centers did not offer transition services, the majority (99%) agreed that a formalized process in transitioning patients to adult care is necessary. A minority (<30%) of respondents stated that they have a written transition policy although 46% have an informal transition process. Designated staff to support transitional care were available in a minority of centers: nurse (35%), physiotherapist (15%), psychologist (15%), social worker (8%), and occupational therapist (2%). The existence of a designated team member to coordinate transition was acknowledged in many centers (64% of respondents) although just 36% use a checklist for young people as part of individualized transitional care.

Conclusion: This survey of European pediatric rheumatology providers regarding transitional care practices demonstrates agreement that transitional care is important, and wide variation in current provision of transition services exists.

Keywords: Transitional care, Rheumatic diseases, Adolescents, Young adults, Chronic disease, Survey method

Juvenile onset rheumatic and musculoskeletal diseases (jRMDs) have high impact on all aspects of the lives of children, young people (YP) and their families. Based on prevalence rates, it is estimated that there are 75,000 children under 16 years old with juvenile idiopathic arthritis (JIA) [1] and about 2500 with systemic lupus erythematosus (SLE) in Europe [2]. These conditions persist into adulthood for many individuals, with continuing disease activity and need for ongoing medication with potential disease-associated morbidity, life-long disability and psychosocial impact [3–5]. During the period of transition, YP with jRMDs have to cope with tremendous physical, emotional and social changes and also with important change in the delivery of clinical care as they move into adult services. Providing transitional care services during this period is necessary to ensure that YP can take control of their health care needs, engage with health care providers and ultimately emerge into adulthood with their optimal function and potential [6, 7]. High-quality, developmentally appropriate healthcare service provision requires the involvement of the YP and the family and also a continuous communication between all persons and provider services involved in transitional care [8].

Whilst the rationale and aims of transitional care in young people with jRMD are clear, organization and delivery of transitional care and what constitutes the ideal service, remains unclear [9, 10]. Previous surveys in the UK,

* Correspondence: minden@drfz.de
[5]Department of Rheumatology and Clinical Immunology, and German Rheumatism Research Centre, Charité University Medicine Berlin, a Leibniz Institute, Berlin, Germany
Full list of author information is available at the end of the article

North America and Europe [11–13] have demonstrated variable provision of care and resources, unmet education and training needs for health care providers, and the need for guidance on the process. This state of play facilitated the drive for the EULAR/PReS working party for transitional care to be set up in 2014 with subsequent published recommendations [14]. The purpose of our survey was to assess as a baseline, the European pediatric rheumatology providers' transition practices and resources in advance of the EULAR/PReS recommendations for transition being widely available.

Methods

European pediatric rheumatologists were invited to complete an anonymized 17-item e-survey assessing their current transition practice. The questionnaire was developed by the authors (DC, LL, HF, LC, KM) after a systematic literature review and critical appraisal of transitional care programs in rheumatology [10] and two face-to-face meetings of the EULAR/PReS working party for transitional care [14]. Items were included that enquired about current transition practices, agreed key elements of transition programs and available resources (see Additional file 1). For comparability reasons, items 1 to 3 were matched with questions of the North-American CARRA survey by Chira et al. [12]. The survey was distributed by the Coordinating Centre of PRINTO (Pediatric Rheumatology INternational Trials Organisation) via SurveyMonkey© to the PRINTO directors of all PRINTO centers in 25 European Union countries. PRINTO is an international network of academic, clinical centers actively engaged in the research/clinical care of children and adolescents with pediatric rheumatic diseases. One reminder was sent to increase the response rate. The questionnaire included items about the transitional care service, staffing, process and resources. The e-survey was piloted by the authors to optimize clarity and ease of administration online.

The study was conducted in cooperation with PRINTO. We did not collect personal identifiable information, such as telephone numbers, names of individuals or institutions, or addresses. The survey respondents voluntarily participated in the study giving their consent to data collection for the purpose of scientific research.

Descriptive statistics are presented and where possible the results were compared with the North American 2014 survey [12] using proportions z test statistical analysis, assuming independence of the two cohorts and $\alpha = 0.01$, given the multiple comparisons being made [15, 16]. The responses of rheumatology centers from different levels of care were compared by the chi-squared test. The 99% CI of the difference between surveys is provided.

Results

Respondent demographics and characteristics

The link to the survey was e-mailed to pediatric rheumatologists (n = 276) from PRINTO centers in 25 European Union countries in April 2016. Of these, 121 responded (44% response rate), representing 115 centers in 22 countries, and thus much of the European Union. Exceptions were Bulgaria, Estonia and Lithuania from whom no responses were received, and Cyprus, Malta and Luxemburg which do not currently have PRINTO centers. The 115 centers who responded correspond to 88% of the PRINTO centers currently listed on the PRINTO-website in those 22 countries. These represent centers with a special interest in research and potentially more likely to have an interest in transition. Most respondents (n = 60; 49%) work in a university-affiliated practice, 39% (n = 47) in a designated children's hospital, 4% (n = 5) in private practice and 7% (n = 8) in other settings.

Transition policy, staff involved, tools and resources

Most (n = 120; 99%) respondents regarded a transition policy as being necessary for good clinical practice. The majority stated that they follow an informal transitional care with only one in four centers having a written transition policy (Table 1). Most rheumatology centers (respondents n = 88; 73%) regularly offer transition services for YP with rheumatic diseases, with a wide range of

Table 1 Current transition policies at the respondents' units

Regarding transition policies….*	Number	Percent
We have a written transition policy, which we follow most of the time.	29	23.9
We have a written transition policy, but we do not follow it most of the time.	4	3.3
We do not have a written transition policy, but follow a fairly standard, informal procedure in transitioning our patients.	56	46.2
We are working on developing a transition policy, but do not yet have one formalized.	21	17.3
We do not have a transition policy, but are interested in developing one.	10	8.2
I do not think that a transition policy is necessary at this point.	1	.8
I have not given it much thought.	0	-

*Percent obtained from 121 affirmative responses

health professionals involved including pediatric and adult rheumatologists and nurses being most common (Table 2). There is a designated staff member who has primary responsibility for coordinating transition process in approximately two thirds of centers (respondents n = 77, 64%); the staff member is often a doctor (n = 60; 78%) or a nurse specialist (n = 14; 18%). A minority (respondents n = 9/119; 7.6%) of transition units receive designated funding or reimbursement for their services, with health insurance companies, grants, pharmaceutical companies, or university as sources.

A minority of centers (respondents n = 44; 36.4%) use a checklist as part of an individualized transition plan that includes the topics shown in Table 3. The majority of these (40/44; 90.9%), agreed that confirmation of the first contact with adult rheumatologist is critical. Other important topics were the importance of knowledge about disease and treatment issues (39/44; 88.6%), as were the encouraging self-management and independent visits by YP without their parents (35/44; 79.5%).

Of note, less than 10% of centers (respondents n = 11) use a specific readiness for transfer instrument, of these, five use a self-developed readiness scale. Other instruments used are the Transition Readiness Assessment Questionnaire (TRAQ), TRANSITION Scale, Ready Steady Go [17].

A minority of respondents (18/121; 15%) reported to use or recommend specific resources for YP and their families. These were mainly websites providing information on juvenile rheumatic diseases, but also on transition aspects, such as the PRINTO website [www.printo.it/pediatric-rheumatology] and the website on transition of the Competence Network Patient Training in Adolescence KomPaS e. V. [www.between-kompas.de]). Furthermore, a minority (38/121; 31%) use information technology to facilitate communication with YP; mainly Short Message Service (SMS), web platforms, email, or apps and the remainder tend to communicate by telephone.

Table 2 Staff involved in the transition services of the respondents' units

Health professionals in the transition service	Number	Percent
Pediatric rheumatologist	92	76.0
Adult rheumatologist	77	63.6
Nurse	42	34.7
Psychologist	18	14.8
Physiotherapist	18	14.8
Social worker	10	8.2
Occupational therapist	3	2.4
Others[a]	11	9.0

[a]Include: internist, medical assistant, expert patients, orthopedic surgeon, gynecologist

Table 3 Components of existing individualized transition plans

Checklist components[a]	Number	Percent
Self-management, patient is seen without parents	35	79.5
Disease and treatment knowledge	39	88.6
Name of disease	39	88.6
Being able to describe disease course	32	72.7
Signs and symptoms of disease flare	33	75.0
Signs and symptoms that require an urgent consultation	30	68.1
Treatment information	40	90.9
Possible side effects of treatment	36	81.8
Health behavior	31	70.4
Risk behavior	29	65.9
Alcohol use, smoking, illegal drugs use	35	79.5
Nutrition	27	61.3
Sports	31	70.4
Mental health	27	61.3
Sexuality, contraception and sexual health	34	77.2
Future plans	33	75.0
Educational achievements	30	68.1
Vocational readiness	22	50.0
Knowledge of support resources	28	63.6
Mobility, living alone, travel	24	54.5
Medical summary available	34	77.2
Knowledge of differences between pediatric and adult rheumatology care	36	81.8
Knowledge about the health system (health insurance, general practitioner/family doctor, health care specialist)	21	47.7
Transfer readiness	28	63.6
First contact to adult rheumatologist	40	90.9

[a]Percent obtained from 44 affirmative responses on having a checklist

Among the rheumatology centers from different models of care, some differences regarding transition care were observed. In comparison to children's hospitals, University affiliated practices provided somewhat less often transition services (69% vs. 78%), had less often a written transition policy (22% vs. 41%) and used significantly less frequently a checklist as part of individualized transitional care (23% vs. 48%, p = 0.011).

Figure 1 is a pragmatic attempt to compare the data from our survey and previous published data from North America [12]. Notably the majority of North American respondents (89%) work in a University-affiliated practice in contrast to 49% of their European colleagues (p < 0.01; 99% CI: 0.29, 0.49). Current transitional care services in Europe and North America show significant differences; the presence of a designated staff member who coordinates transition (64% in Europe vs 30% in North America; p < 0.01; 99% CI: -0.45, −0.22) and the availability of a

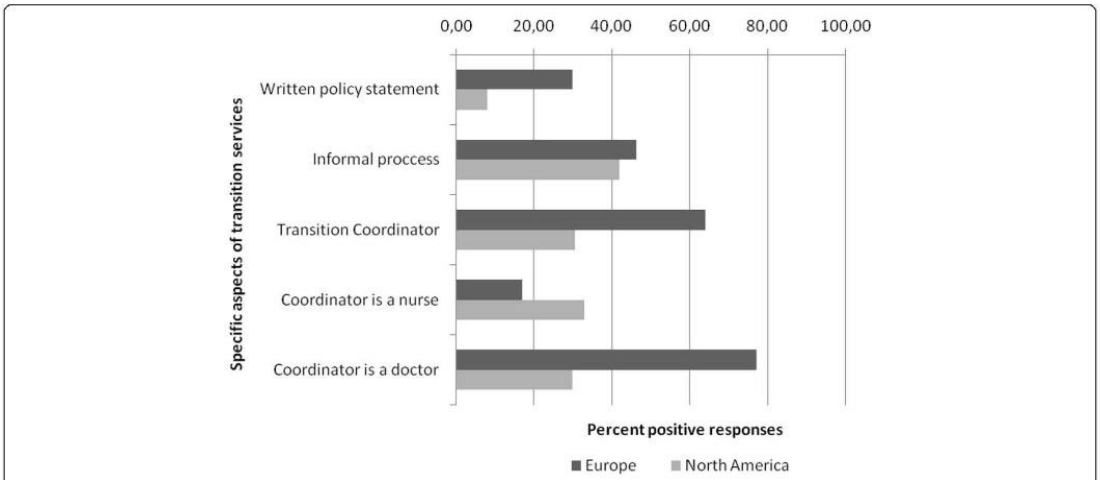

Fig. 1 Comparison of transitional care services offered by European and Childhood Arthritis and Rheumatology Research Alliance (CARRA) pediatric rheumatology providers

written transition policy (26% in Europe vs 8% in North America; $p < 0.001$; 99% CI: -0.26, −0.08). Nevertheless, the percent of informal transition process is similar across surveys (46% vs. 42%; non-significant).

Discussion

This is the first survey to focus on the current practices (rather than solely attitudes or opinions) in transitional care among European pediatric rheumatology providers. As in other chronic diseases that start in childhood, YP with jRMD need specific care during the period of transition from pediatric services to the adult specialty care setting. However, research about current practice and the implementation of transition recommendations are scarce and almost half of the YP with jRMD do not make a successful transfer to adult rheumatology and are, therefore, at increased risk of unfavorable outcomes [5, 18, 19].

Recently, EULAR/PReS recommendations were issued for transitional care throughout Europe [14]. These recommendations were published after the e-survey was conducted and so our data serve as useful benchmark of current practice. The recommendations highlight the need for a written transition policy, and yet less than a third of respondents in our survey indicated that their department had a formal transition program; an 'informal approach' was reported to be more common although further information was not given in the survey. It is hoped that the use of the recommendations and a consistent approach to the transition program is more likely to address the structure, assessments of patient readiness and service outcomes. We note that only a small percentage of transition clinics receive funding or reimbursement for the service and it is possible that one barrier to implement more ambitious transition programs may be insufficient resources. Interestingly, the EULAR/PReS recommendations include the need to secure funding for provision of transitional care, in order to avoid some of the major barriers as highlighted by the North American survey and a frequent shortcoming in Europe [14].

The presence of a transition coordinator is another key element of transition and one of the EULAR/PReS recommendations; the aim being to ensure implementation and evaluation of the transition program and improving communication between health professionals, YP and families [8, 14]. Most respondents of the survey have a designated staff member as transition coordinator. However, in contrast to the recommendations, emphasizing the important role of allied health professionals in transition coordination, we note that the designated staff member is often a doctor rather than a nurse, youth worker or other professional. The EULAR/PReS recommendations do not specify whether the coordinator is a doctor or allied health professional but that the individual is clearly identified within the team and has the appropriate skills to facilitate liaison between the pediatric and the adult care teams [14].

Information and education of YP about their disease, treatments and promotion of independent visits to clinic without parents are ideal components of an individualized transition plan. However, it is important to include systematically other important areas of adolescence health, such as risk behavior, substance use, sports, nutrition or sexuality. The HEADSS assessment (Home, Education/Employment, peer group, Activities, Drugs, Sexuality, Suicide/depression) was developed [20] and has since been successfully used to facilitate communication with YP and

as guide to cover all topics pertinent to young people in the transition process. However, to address all relevant transition issues, it might be necessary to have a multidisciplinary team (i.e., psychologist to handle mental health or physiotherapist to advice about sports and routine exercises). This approach is clearly recommended in the EULAR/PReS document [14]. Our survey revealed that currently only a minority of centers offers multidisciplinary transitional care services; 35% of the centers have a nurse, 15% a psychologist and 8% a social worker available for addressing transition-relevant issues.

Our survey did not specifically address timing of transfer. However, it should be noted that only 9.5% use a readiness instrument to validate transfer, reflecting the informal approach to transition in the majority of centers. This issue was heavily discussed in the development of the EULAR/PReS recommendations, as some countries had a designated age for transfer in all cases and independently of YP readiness. Ultimately the EULAR/PReS recommendations propose that age per se is not specifically used as the trigger for timing of the transfer. Conversely, age is an important indicator for the initiation of transitional care; namely that the transition process should start as early as possible [14]. The recommendations also propose strategies to facilitate transfer including communication with adult services before actual transfer with relevant information about the YP or shared clinics between pediatric and adult health care professionals. In this way, the timing of transfer could be more flexible, and could be deferred until the disease is stable and/or the transition team consider the patient to be "ready".

Where possible and with the questions that can be compared, we were able to comment on differences between European and North American respondents. European providers more often do have a transition coordinator (often a doctor) and are more likely to use a written transition policy statement. In both surveys, respondents manifest a strong desire for rheumatology-specific guidelines for transition [12] and for these to be implemented across pediatric, adolescent and adult rheumatology health care settings.

In summary, this survey has demonstrated limitations of existing transition practices and paucity of resources. Nonetheless there is a strong commitment within the rheumatology communities (both adult and pediatric), to improve existing transitional care provision. The EULAR/PReS recommendations are therefore timely and important as a much needed catalyst for change within the rheumatology community.

Additional file

Additional file 1: Questionnaire on transition practices. (DOC 36 kb)

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Availability of data and material
The data of this survey are available from the last author on reasonable request.

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Authors' contributions
All authors were involved in drafting the article and approved the final version to be published. Survey conception and design: DC, LL, HF, LC, KM. Acquisition of data: KM. Analysis and interpretation of data: DC, LL, HF, LC, KM.

Competing interests
The authors declare that they have no competing interests.

Consent for publication
Not applicable.

Ethics approval and consent to participate
Not applicable.

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Author details
[1]Paediatric Rheumatology Unit, Hospital Infantil Universitario "Niño Jesús", Madrid, Spain. [2]IDISSC, Hospital Clínico San Carlos; Health Sciences, Universidad Camilo José Cela, Madrid, Spain. [3]Musculoskeletal Research Group, Institute Cellular Medicine, Newcastle University, and Great North Children's Hospital, Newcastle Hospitals NHS Foundation Trust, Newcastle upon Tyne, UK. [4]Instituto de Salud Musculoesquelética, Madrid, Spain. [5]Department of Rheumatology and Clinical Immunology, and German Rheumatism Research Centre, Charité University Medicine Berlin, a Leibniz Institute, Berlin, Germany.

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EULAR/PReS standards and recommendations for the transitional care of young people with juvenile-onset rheumatic diseases

Helen E Foster,[1] Kirsten Minden,[2,3] Daniel Clemente,[4] Leticia Leon,[5,6] Janet E McDonagh,[7] Sylvia Kamphuis,[8] Karin Berggren,[9] Philomine van Pelt,[10] Carine Wouters,[11] Jennifer Waite-Jones,[12] Rachel Tattersall,[13] Ruth Wyllie,[14] Simon R Stones,[15] Alberto Martini,[16] Tamas Constantin,[17] Susanne Schalm,[18] Berna Fidanci,[19] Burak Erer,[20] Erkan Dermikaya,[21] Seza Ozen,[22] Loreto Carmona[23]

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For numbered affiliations see end of article.

Correspondence to
Dr Leticia Leon, Instituto de Investigación Sanitaria del Hospital Clínico San Carlos (IDISSC), Hospital Clínico San Carlos, Martin Lagos s/n, Madrid 28040, Spain; lleon.hcsc@salud.madrid.org

HEF and KM contributed equally.

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ABSTRACT

To develop standards and recommendations for transitional care for young people (YP) with juvenile-onset rheumatic and musculoskeletal diseases (jRMD). The consensus process involved the following: (1) establishing an international expert panel to include patients and representatives from multidisciplinary teams in adult and paediatric rheumatology; (2) a systematic review of published models of transitional care in jRMDs, potential standards and recommendations, strategies for implementation and tools to evaluate services and outcomes; (3) setting the framework, developing the process map and generating a first draft of standards and recommendations; (4) further iteration of recommendations; (5) establishing consensus recommendations with Delphi methodology and (6) establishing standards and quality indicators. The final consensus derived 12 specific recommendations for YP with jRMD focused on transitional care. These included: high-quality, multidisciplinary care starting in early adolescence; the integral role of a transition co-ordinator; transition policies and protocols; efficient communications; transfer documentation; an open electronic-based platform to access resources; appropriate training for paediatric and adult healthcare teams; secure funding to continue treatments and services into adult rheumatology and the need for increased evidence to inform best practice. These consensus-based recommendations inform strategies to reach optimal outcomes in transitional care for YP with jRMD based on available evidence and expert opinion. They need to be implemented in the context of individual countries, healthcare systems and regulatory frameworks.

INTRODUCTION

Transitional care, as defined by the Society for Adolescent Medicine, is "the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centred to adult-oriented healthcare systems".[1] Transition focuses on the administrative event of transfer of care between paediatric and adult providers; transition encompasses the process by which young people (YP) acquire skills and access resources to ensure that their physical, psychosocial, educational and vocational needs are met during transition to adulthood.[2] Adolescence and young adulthood reflect an important and unique developmental period for all YP who need education, support, guidance and planning to prepare them to be appropriately responsible and accountable for their own health and well-being as adults.[3] The same principle applies to YP with chronic illnesses (including juvenile-onset rheumatic and musculoskeletal diseases (jRMDs)), who need to acquire additional skills to independently manage their chronic illness. The case of need for transition is well described and transitional care aims to provide support and guidance so that YP can acquire the necessary skills and knowledge required to be independent, empowered and responsible adults.[4–7]

The course of jRMDs often continues into adulthood; according to population-based inception cohort studies, approximately half of YP with jRMDs enter adulthood with active disease, or develop flares of disease as adults. Many YP require ongoing and often long-term treatment with complex immunosuppressive regimes.[8–10] Disease-related sequelae are still observed, although with modern approaches to management, many YP are transferring to adult care in clinical remission albeit on medication.[11–13] All YP with jRMDs are, in principle, at significant risk of disability, early morbidity and limitations in participation later in life.[14 15] These YP need continuous and developmentally appropriate care during and beyond adolescence to ensure optimal functioning in adulthood. However, the literature informs us that currently up to half of the YP do not make a successful transfer to adult rheumatology and are therefore at particular risk of unfavourable outcomes.[16–18]

The importance of transitional care in YP with jRMD has been increasingly acknowledged.[19 20] There is evidence regarding 'best practice' for transitional care, with emphasis on a holistic 'life course' approach to care.[21–23] Several healthcare institutions, specialties and disease-specific subgroups have developed and implemented transition programmes.[24–34] However, there is lack of clarity regarding the impact of transition programme on outcomes, and indeed, variance in what the outcomes should be.[35–40] A first consensus-based proposal regarding outcome indicators for successful healthcare transition was recently made by an

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international group of interdisciplinary healthcare professionals, patients and their families.[41] With rheumatology, there are significant gaps in current delivery of transition services and these include the unmet training needs for healthcare professionals in adolescent health and transitional care (resulting in lack of understanding and appreciation of the needs of YP), lack of transition readiness of YP (and/or of their parents/carers) and lack of robust quality indicators or cost-effective strategies.[2 36 42–48]

Despite the limitations of the existing programmes, the gaps in knowledge and the paucity of resources, there is nonetheless commitment within the rheumatology communities (both adult and paediatric), to improve existing transitional care services. There is a strong desire for rheumatology-specific guidelines for transition[48] and for these to be implemented across paediatric, adolescent and adult rheumatology healthcare settings.

Objectives, scope, users and overarching principles

The objective of the present initiative was to develop recommendations and standards for transitional care for YP with jRMDs, spanning ages from early adolescence (defined as 10–13 years), mid adolescence (14–16 years), late adolescence (17–19 years) to young adulthood (20–24 years).[49 50] These recommendations and standards are to be used to guide service development, benchmark the quality of transition services and be used by patient organisations to enhance patient expectations of care. We acknowledge that their implementation into clinical practice will be challenging and likely to be facilitated by stratification into 'essential' and 'ideal' components—*essential* defined as the *minimum* standards below which care would be deemed unacceptable and ideal being the standard that is regarded as excellent 'optimal' care.

The purpose of these recommendations and standards is to increase the profile of transition, optimise delivery of transitional care and improve patient experience within rheumatology across European countries. Specifically, the objectives are:

- to ensure youth friendly and developmentally appropriate care,
- to improve physical, psychological, social, vocational and illness-related outcomes of YP with jRMDs,
- to facilitate continuity of care within adult rheumatology,
- to promote evidence-based practice in transitional care,
- to facilitate clinical networks of healthcare professionals (paediatric and adult), who are engaged, interested and trained in the care of YP.

The scope of these recommendations and standards refers to all persons involved in the care of YP with jRMDs that continue into adulthood including, but not restricted to, those in box 1.

Although these recommendations and standards are related to the specific needs of YP with jRMDs, our expert panel endorse the American Academy of Pediatrics, the American Academy of Family Physicians and the American College of Physicians-American Society of Internal Medicine consensus statement on healthcare transitions for YP with special healthcare needs,[51 52] position papers of the Society for Adolescent Medicine,[1 53–55] the Canadian Paediatric Society,[56] the Royal Australasian College of Physicians[57] and the WHO definition of adolescent-friendly health services.[58 59] In addition, we emphasise key components integral to these recommendations and standards, namely the importance of a YP focus, multidisciplinary approach with equity of access, quality of care and flexibility; the latter acknowledging both the heterogeneity of YP development and potential impact of chronic illness.

Box 1 Individuals to whom these recommendations may prove useful

Adult and paediatric rheumatologists
Other healthcare professionals:
- nurses
- physiotherapists
- occupational therapists
- social workers
- psychologists
- youth workers
- primary care and other specialists (eg, ophthalmology, dermatology, nephrology, orthopaedics) involved in share care and clinical networks

General practitioners and healthcare professionals working in primary care
Young people with rheumatic and musculoskeletal disease (RMD) and other multisystem disease, their families and peers
Professional groups or societies working with young people with RMDs
Academics involved in adolescent and young adult health research
In addition, these standards and recommendations are intended to be useful to the remit of
- educational and vocational services
- employers and careers advisory services
- charities and support organisations
- funders of clinical services
- health policy makers
- research funding bodies
- organisers of disease and/or drug registers, and cohort studies

METHODS

The consensus process underwent the following stages: (1) establishing an international expert panel to include patients and representatives from multidisciplinary teams (MDTs) in adult and paediatric rheumatology; (2) a systematic literature review; (3) setting the framework, developing the process map and generating a first draft of standards and recommendations; (4) further iteration of recommendations; (5) developing consensus recommendations with Delphi methodology and (6) establishing standards and quality indicators, as suggested by the European League Against Rheumatism (EULAR) Standard Operating Procedures.[60]

The project convenors (HF, KM, LC) liaised to appoint clinical fellows (DC, LL) to work on the project. They then convened an expert multidisciplinary panel from adult and paediatric rheumatology across Europe (doctors and allied health professionals with interest in transitional care) and patient representatives (YP with jRMDs invited from existing patient groups). The aim was for the panel to reflect the diversity of Europe (namely geography, healthcare systems and cultures).

A systematic literature review of existing models of transitional care in jRMD was performed, with emphasis on potential recommendations, standards, strategies for implementation and tools to evaluate services and outcomes.[35]

The first 'face-to-face' meeting of the expert panel discussed results of the systematic review and agreed the following:

(i) purpose of the project, timelines, roles and planned outputs;

(ii) the 'process map' of transitional care using MindManager software;
(iii) a draft proposal of recommendations and standards relating to different elements of the process map; the proposed list was circulated after the meeting by email to the expert group and further feedback requested.

A second 'face-to-face' meeting further refined the recommendations and standards based on feedback from the group. Appropriate quality indicators were also suggested for each recommendation and standard.

The recommendations and standards were then listed as statements. A wider audience of a total of 195 adult and paediatric rheumatology clinicians (doctors and allied health professionals) were then invited to take part in an e-survey and give opinion of their level of agreement with each statement. The e-survey was disseminated through email lists held by professional groups (such as Paediatric Rheumatology European Society (PReS), EULAR and rheumatology societies in different countries). All responses were anonymised. Participants were asked for their level of agreement with each statement (using a 10-point Likert scale, with 0=no agreement through to 10=total agreement) and a ranking exercise to identify 'minimal' and 'optimal' standards for each recommendation. The target number of respondents for the e-survey was 100 and the level of agreement set at 80% for acceptance (lower levels were to be then discussed by the expert panel, with further iterations of the statements proposed and then a second e-survey, if needed, to be disseminated). Once agreement was reached, the methodologist (LC), together with the clinical fellows (DC, LL), graded the level of evidence for each recommendation based on the Oxford Levels of Evidence, 2011 (available at http://www.cebm.net/index.aspx?o=5653) and assigned relevant quality indicators where appropriate.

RESULTS

The recommendations and standards

The recommendations and standards, reported as 'minimal/essential' and 'ideal/optimal' levels of care and quality indicators are listed below; they are also presented in table format with the level of evidence and agreement reached (table 1).

(1) YP with jRMDs should have access to high-quality, co-ordinated transitional care, delivered through partnership with healthcare professionals, YP and their families, to address their needs on an individual basis.

High-quality care means holistic care (which covers medical, psychosocial, educational and vocational aspects; see box 2) with a multidisciplinary approach and is based upon regular assessments of the disease status in the context of developmental stage, life events and personal aspirations of YP. This care needs to be 'future-focussed', albeit not limited to young adulthood, in order to ensure optimal well-being.[19 23]

Ideal: provision of a broad set of healthcare services led by providers who have specialist knowledge about jRMDs and adolescent health; essential: care providers who refer to other agencies and services who can appropriately assist with transition issues.

(2) The transition process should start as early as possible; in early adolescence or directly after the diagnosis in adolescent-onset disease.

Specifically for childhood-onset diseases, the transition process should start by early adolescence (11 years) (ideal) or 14 years at the latest (essential) in order to allow the development of the necessary self-care skills and optimise educational and vocational outcomes.[63 64] For YP who are diagnosed over the age of 14 years, the transition process should start at the time of diagnosis with the skills and support for transition built-up over time. The transition process or joint care programmes should enclose early adulthood, because young adults continue to have difficulties making effective linkages with adult care. Brain development continues and risk behaviours remain and may increase in the third decade, which have to be considered by proactive and preventive care.[65]

(3) There must be 'direct' communication between the key participants (and as a minimum, to include the YP, parent/carer and a member of each of the paediatric and adult rheumatologist teams) during the process of transition. Before and after the actual transfer, there should be 'direct' contacts between paediatric and adult rheumatologist teams.

A network of adult rheumatologists interested, engaged with and trained in adolescent rheumatology, must be identified and known to the paediatric rheumatology team. The network

Table 1 Recommendations, standards and proposed quality indicators on transitional care for young persons with RMD

Recommendations	LOE	GR	MA
1. YP with RMD should have access to high-quality, co-ordinated transitional care, delivered through partnership with healthcare professionals, YP and their families, to address needs on an individual basis	5	D	9.6
2. The transition process should start as early as possible; in early adolescence or directly after the diagnosis in adolescent-onset disease	2b	B	8.3
3. There must be 'direct' communication between the key participants (and as a minimum, to include the YP, parent/carer, and a member each of the paediatric and adult rheumatologist teams) during the process of transition. Before and after the actual transfer, there should be 'direct' contacts between paediatric and adult rheumatologist teams	5	D	9.3
4. Individual transition processes and progress should be carefully documented in the medical records and planned with YP and their families	5	D	9.2
5. Every rheumatology service and clinical network—paediatric and adult—must have a written, agreed and regularly updated transition policy	5	D	8.9
6. There should be clear written description of the MDT involved in transitional care, locally and in the clinical network. The MDT should include a designated transition co-ordinator	5	D	8.7
7. Transition services must be YP focused, be developmentally appropriate and address the complexity of YP development	5	D	9.4
8. There must be a transfer document	5	D	9.4
9. Healthcare teams involved in transition and adolescent-young adult care must have appropriate training in generic adolescent care and childhood-onset RMD	5	D	9.5
10. There must be secure funding for dedicated resources to provide uninterrupted clinical care and transition services for YP entering adult care	5	D	9.4
11. There must be a freely accessible electronic-based platform to host the recommendations, standards and resources for transitional care	5	D	9.4
12. Increased evidence-based knowledge and practice is needed to improve outcomes for YP with childhood-onset RMD	5	D	8.5

GR, grade of recommendation; LOE, level of evidence; MA, mean agreement (0–10); MDT, multidisciplinary team; YP, young people.

Box 2 Aspects considered as part of holistic care

Medical aspects:
- identification of medical needs, addressing any issues
- ensuring continuity of provision of high-quality care
- providing generic and disease-specific information
- health promotion, anticipatory guidance
- health behaviour (eg, health literacy, experimentation and risk behaviour), negotiating most appropriate ways to ensure adherence to treatment
- knowledge and skills in areas listed above

Psychosocial aspects:
- identifying individual needs, risk and protective factors (eg, Home, Education, Activities, Drugs, Sex, Suicide (HEADSS))[61 62]
- providing support or referring young people to specific agencies
- ensuring a social life that is equivalent to those of peers
- ensuring support to cope with disease/treatment
- providing advice and/or additional sources of support
- promoting skills in assertiveness, resilience, self-care, self-determination and self-advocacy

Educational and vocational aspects:
- addressing future career prospects
- developing skills in disclosure
- support in preparing for work readiness
- informing about where to get information (recommend: career advisors, appropriate agencies, charity websites)
- addressing work experience and encouraging young people to gain relevant experience
- offer appropriate information, support and advice (support groups, volunteer services)
- liaisons with educational institutions
- informing about rights and obligations, benefits and opportunities to adapt working (place, time)

should work within agreed pathways to facilitate transition and expedite early, active planning of transition. Ideally, there should be a combined meeting between the young person and his or her family, the paediatric and adult healthcare provider.[66–68] As a minimum, there should be at least two 'direct' contacts by telephone or email between the paediatric and adult rheumatology team (and documented in a written communication); one before and one after the transfer. Copies of written communications are to be made available to YP and families. Online supplementary table contains suggested guidelines regarding the content and format of communications at different stages of transitional care.

(4) Individual transition processes and progress should be carefully documented in the medical records and planned with YP and their families.

Documentation should support the YP engagement and self-management skills, resilience and readiness for transfer.[69–73] This documentation should be tailored to local services, shared with the YP and contribute to the medical summary. The inclusion of sensitive or confidential information (eg, abortion, mental health problems) should be discussed with the YP. Ideally, there is a specific written individual transition plan,[26] which can be derived from a transition plan or passport example, such as http://www.uhs.nhs.uk/Media/Controlleddocuments/Patientinformation/Childhealth/ReadySteadyGo/Ready-Steady-Go-Transition-plan.pdf.[21] As a minimum, the existence of a transitional care process has to be documented in the medical records. Additional resources are listed in Supplementary material.

(5) Every rheumatology service and clinical network—paediatric and adult—must have a written, agreed and regularly updated transition policy.

Policies and protocols should be agreed with all major stakeholders, including YP, families and all healthcare professionals and as equal partners;[27 33 35] it is important to stress the need to include all specialists (and not just in rheumatology) and primary care physician(s) who are involved in the clinical care of YP with jRMD[28] Hospital or institutional managers will have to agree to these policies to facilitate appropriate resources to support their implementation within the clinical departments. As a minimum, there must be a transition policy and the documents should be updated at least every 5 years. It is acknowledged that there is need for flexibility of the arrangements in transition policy and care pathways at a network level.

(6) There should be clear written description of the MDT involved in transitional care, locally and in the clinical network. The MDT should include a designated transition co-ordinator.

The team for transitional care should reflect the multidisciplinary approach, that is, doctors and other health professionals, such as nurses, physical therapists, psychologists, occupational therapists and youth or social workers.[28 74 75] It is recognised that certain roles within transition are likely to be addressed by different members of the MDT.[76] In addition, it is acknowledged that the composition of MDTs is variable and that some members may have more than one role. Ideally, there is personal continuity in the health professionals within the MDT providing care.[43] This provision of roles and services may be shared with other specialist services and not devoted solely to jRMDs. There should be a nominated and identified member of the MDT, who is responsible as transition co-ordinator (essential). This person can be a nurse or other health professional and should liaise between adult and paediatric teams to ensure the co-ordination of care, facilitation of communication and implementation of the transitional care plan including transfer.[35]

(7) Transition services must be YP focused, be developmentally appropriate and address the complexity of YP development.

Components of YP focused care need to include accessibility to specialised healthcare, staff attitudes, communication, medical competency, guideline-driven care, age appropriate environments and youth involvement in healthcare.[77] Ideally, there should be a care facility that is truly adolescent-friendly staffed by professionals with expertise in adolescent care. The care should be organised to minimise the frequency of appointments and interruption to the daily life of YP. The aim of the consultations with the MDT is to enable YP to take lead role instead of the parent/carer(s), while also supporting the parent/carer(s) in their changing roles.[43 78 79]

YP with jRMDs should have access to peer discussion and support through advisory group(s) and charity networks. Signposting to such groups and networks is the responsibility of the clinic where the YP attends. As a minimum, transitional care services should be led by staff with expertise and training in adolescent and young adult healthcare.

(8) There must be a transfer document.

The format and content of transfer documents should be agreed by paediatric and adult teams and with patient input where possible. The transfer document should include, as a minimum, a medical summary with the diagnosis, any comorbidities, vaccinations, any complications of disease or treatments, the professionals involved in care, current and

previous treatments (with reasons for changing treatments and any adverse events). Ideally, it should also include: (i) psychosocial aspects and educational/vocational status at the time of transfer and (ii) a report on self-management skills, to include readiness for transfer and procedural pain management strategies (eg, for joint injections with or without general anaesthesia).[80]

This transfer document may also include contributions from members of the MDT as appropriate and if relevant. Any confidential information or sensitive information to be included in the transfer document should be discussed with the YP; if needed, this should be included in a separate letter to the adult rheumatologist and be written ahead of the first consultation with adult rheumatology. Copies of the transfer document should be available to YP themselves in an easy read format if appropriate, and to all healthcare providers, including primary care, involved in the young person's care.[81]

(9) Healthcare teams involved in transition and adolescent-young adult care must have appropriate training in generic adolescent health and childhood-onset RMD.

All members of the clinical MDT (from adult and paediatric rheumatology) engaged in transition are to have training on adolescent health and the process of transition.[46 48] As a minimum, the key training components to be covered are: (1) jRMDs (presentation in childhood, knowledge and approaches to management), (2) adolescence health and the impact on jRMDs, (3) skills and knowledge to address emotional, mental health and social issues, (4) promotion of healthy lifestyle and generic health issues, (5) promotion of self-management and shared decision making, (6) communication skills with YP and their parent/carer(s).

These skills and knowledge can be acquired through different ways of learning (eg, to include clinical experience, e-learning and practical workshops) and should be a component of continuous professional development. There are many courses and e-learning opportunities available, such as the EULAR/PReS On-line Course in Paediatric Rheumatology (http://www.eular.org/edu_online_course_paediatric.cfm), the European Training in Effective Adolescent Care and Health (http://www.unil.ch/euteach/en/home.html) or the UK Adolescent Health Project (http://www.e-lfh.org.uk). Reciprocal periods of training for members of the adult and paediatric rheumatology teams are advocated (ideal).

(10) There must be secure funding for dedicated resources to provide uninterrupted clinical care and transition services for YP entering adult care.

The following are regarded as essential for transitional care programmes:

(i) The funding and supporting resources for care should be dependent on clinical need and should not be interrupted on sole grounds on the age of the patient. Conversely, the presence or absence of resources should not define the timing of transfer.
(ii) Funding of (biological and other) therapies should continue if clinically indicated irrespective of patient age and transfer to adult care.
(iii) Funding of the paediatric and adult MDTs involved in the transitional care is needed.
(iv) The transition co-ordinator role and administration support for clinical networks must be funded.
(v) The training for the MDT along with continuous professional development support must be funded.
(vi) The importance of adequate administrative support is likely to be a determinant of successful transfer.[45 48 81]

(11) There must be a freely accessible electronic-based platform to host the recommendations, standards and resources for transitional care.

An open resource e-platform (essential), such as the EULAR/PReS website, to host resources to support the transition process, staff training and patient resources, would facilitate setting up of new and further development of existing transition services. All stakeholders must have access to these resources, including YP with jRMDs, their families and healthcare professionals in hospital, primary and community care involved. The resources must be endorsed (essential) by professional bodies such as EULAR/PReS, consumer groups and charities in the respective countries.

(12) Increased evidence-based knowledge and practice is needed to improve outcomes for YP with childhood-onset RMD.

The expert group recognised the need for a greater evidence base to inform best practice, the best metrics for measuring 'success' and 'outcome' of transitional care programmes and the impact of such programmes on YP with jRMDs.[7 37 38] The agenda for research needs to include:

(i) the transition programme evaluation as a complex intervention,[75]
(ii) the effectiveness of the transition process[35] and how this can be measured,
(iii) the timeliness of interventions and validation of readiness tools,[69–73 82]
(iv) the outcome measures of transition,[37–39 41 83]
(v) predictors for transition outcomes,
(vi) cohort studies and registries from paediatric rheumatology to extend into adult life,
(vii) an agreed 'core transition dataset' for routine practice in paediatric and adult rheumatology centres to inform and foster future research initiatives, facilitate a standardised approach in transitional care and enable comparative assessment of care across Europe.

Standards and quality indicators

The expert group concluded that there is a limited evidence base for outcomes of transition and for YP rheumatology services.[35] The expert group proposed, before the publication of the previous studies, key quality indicators to measure service delivery and to inform the research agenda. Online supplementary table shows a list of standards and the agreed quality indicators.

In 2015, Suris and Akre[80] co-ordinated an international consensus on key elements and one indicator of a good transition. In most instances, such consensus underscored items related to co-ordination and communication as basis for good partnership between paediatric and adult providers. Another initiative, developed during our project, obtained a clinical practice-benchmark tool for transition to adult care in the UK through a process of mapping.[84] There is no single outcome of 'successful' transition and potential indicators need to include clinical parameters (disease activity and status), patient (and family) experience (of care), psychosocial, educational and vocational status, quality of life measures, participation in adult life, engagement and attendance in adult healthcare, adherence to treatment and achievement of young adult developmental tasks. A recent taskforce identified, by Delphi methodology, outcomes of importance.[41] These included individual outcomes (quality of life, understanding the characteristics of conditions and complications, knowledge of medication, self-management, adherence to medication and understanding health insurance), health services outcomes (attending medical appointments, having a

medical home and avoidance of unnecessary hospitalisation) and a social outcome (having a social network). Measures need to be valid, reproducible and relevant.

DISCUSSION

This PReS/EULAR taskforce has developed the first international set of recommendations and standards for transitional care of YP with jRMDs; the aim being to facilitate high-quality models of care for new and existing services, inform strategies for evaluation and define a research agenda. These were produced before the emergence of the 2016 National Institute for Health and Care Excellence (NICE) guidance on Transition in the UK (http://www.nice.org.uk/guidance/ng43) but reassuringly, are consistent NICE recommendations. Our methodology permitted critical appraisal of published models of care and incorporated opinion from a diverse expert specialists group including YP.[35] Our recommendations and standards set out the 'essential/minimal' and 'ideal/optimal' components of transitional care and we anticipate that such stratification will be helpful to benchmark services and facilitate implementation and evaluation.

The recommendations focus on transitional rheumatology care, however, they comprise components of high-quality transitional care derived from policy documents, guidelines relating to transitional care and the adolescent health literature. This underlines that, overall, most key elements of transitional care are generic. That is also reflected in the key elements for successful transition, which were published by Suris and Akre[80] after the recommendations given here were agreed on. Six elements were regarded as being essential by more than 70% of an international panel, of which two relate to establishing a good partnership between paediatric and adult professionals and the shared responsibility of transitional care. All six essential elements are included in the recommendations given here, which highlight the need for teams to work effectively together and engagement of different care providers within clinical networks. Addressing the challenges of 'joined up' working across paediatric and adult rheumatology and within clinical networks has also been highlighted by others.[85] Our recommendations, in agreement with other recent taskforces,[80 84] emphasise the importance of identifying key individuals, the integral role of YP and families, written communication, agreed policy, training and clarity of roles within teams. Therewith, they focus on process areas that are most in need of improvement according to care providers and consumers, such as co-ordination, guidelines, protocols and communication.[66] It is apparent that a transitional care pathway for YP with jRMDs can be implemented with a motivated healthcare team, the reorganisation of their existing work practice and available resources.[29 31] The transitional care MDT also needs specific training in adolescent medicine and adequate capacity to enable the transition care co-ordinator role to function.[46 48] Transition is resource consuming. The expert panel stresses the fact that without sufficient funding or reimbursement of the specific interventions, transitional care services cannot become a normal part of healthcare for YP. Funding is needed for specific service provision and to ensure continuity of clinical care and access to medicines after transfer to adult care based on clinical need rather than age of the patient of the provider. There are promising examples in the UK or Germany, where the provision of transitional care services as part of clinical practice has been funded by the government or statutory health insurance companies within defined programmes.[32] The funding for continuity of clinical care and access to medicines after transfer to adult care has to be addressed and based on clinical need rather than age of the patient of the provider. Given the importance of transition for the many YP who transfer to adult rheumatology, we strongly suggest that transitional care is included in all PReS and EULAR activities to raise awareness, promote access to training and improve skills and knowledge among all adult rheumatology teams.

Quality indicators and outcomes of transitional care are proposed. These transitional care outcomes are similar to most of 10 prioritised outcomes identified by a task force from the Health Care Transition Research Consortium[41] and may allow researchers to conduct focused evaluations of current processes and more detailed evaluations of interventions.

We acknowledge differences between countries in how transition may be organised within different healthcare systems.[66 86–88] Our recommendations are intended to be useful, widely applicable and promote transitional care. Although transitional care has received much attention in the child health community, little government attention has been paid to this complex health system issue. An analysis of policy profiles of paediatric-to-adult care transitions in six European countries revealed that four had currently no transition policies or strategies.[88] The overarching principles seek to promote transferability to different contexts, be compliant with national regulatory guidance and facilitate local teams to work together with responsibility and accountability for services to be suitable for local needs. Our approach is similar to that proposed by others; the Spanish consensus for transition management in patients with jRMDs[89] reported a framework with similar recommendations albeit with more practical details suited to the Spanish healthcare system.

Transition is a time-variable process that prepares YP with jRMDs to take responsibilities for their lives and also their health issues. This process is critical in order to facilitate the actual transfer to adult care. Transition is therefore a complex process with many variables involved and the panel required considerable dialogue and indeed compromise to agree the aims, framework and process map to address such complexity. However, as the project evolved and moved forward, harmonisation became apparent and ultimately one round of Delphi was adequate to achieve high agreement.

It is clear that there are many unanswered questions in transitional care. Our recommendations highlight the need for improved evidence base to inform models of care, identify relevant outcome measures and the cost-effectiveness of transitional care programmes as a complex intervention. Much work is yet to be done, but it is important to identify and ultimately deliver 'best' care for YP with jRMDs and their families, to facilitate optimal physical, psychosocial and quality of life outcomes within adulthood.

Author affiliations
[1]Newcastle University, Institute of Cellular Medicine (Rheumatology), Newcastle, UK
[2]Children's University hospital Charité, Campus Virchow, SPZ, Berlin, Germany
[3]Epidemiology Unit, German Rheumatism Research Centre, Berlin, Germany
[4]Paediatric Rheumatology Unit, Hospital Infantil Universitario "Niño Jesús", Madrid, Spain
[5]Hospital Cínico San Carlos, Instituto de Investigación Sanitaria del Hospital Clínico San Carlos (IDISSC), Madrid, Madrid, Spain
[6]Health Sciences, Universidad Camilo José Cela, Madrid, Madrid, Spain
[7]University of Manchester, Centre for Musculoskeletal Research, Manchester, Manchester, UK
[8]Immunology and Infectiology, Erasmus MC—Sophia, Rotterdam, The Netherlands
[9]Karolinska Universitetssjukhuset, Stockholm, Sweden
[10]Department of Paediatrics/Paediatric Rheumatology, Erasmus MC Sophia Children's Hospital Rotterdam, Rotterdam, The Netherlands
[11]Pediatric Immunology, University Hospital Gasthuisberg, Leuven, Belgium
[12]University of Leeds, School of Healthcare, Leeds, UK

[13]University of Sheffield, School of Health and Related Research, Sheffield, Sheffield, UK
[14]Paediatric Rheumatology, Newcastle upon Tyne Hospitals NHS Foundation Trust, Newcastle upon Tyne, Newcastle upon Tyne, UK
[15]London, UK
[16]Pediatria II, Universita degli Studi di Genova Scuola di Scienze Mediche e Farmaceutiche, Genova, Liguria, Italy
[17]Department of Pediatrics, Semmelweis Egyetem, Budapest, Hungary
[18]Transitionssprechstunde am Dr. von Haunerschen Kinderspital, Ludwig-Maximilians-Universität München, Munchen, Germany
[19]Gulhane Military Medical Academy, School of Nursing, Ankara, Turkey, Ankara, Turkey
[20]Division of Rheumatology, Department of Internal Medicine, Istanbul University, Istanbul Faculty of Medicine, Istanbul, Turkey
[21]Gulhane Military Medical Faculty, Pediatric Rheumatology Unit, FMF Arthritis Vasculitis and Orphan Disease Research in Pediatric Rheumatology (FAVOR), Etlik, Ankara 06018, Turkey, Ankara, Turkey
[22]Pediatric Rheumatology, Hacettepe University Medical Faculty, Ankara, Turkey
[23]Instituto de Salud Musculoesquelética, Madrid, Spain

Contributors HeEF, KM, DC, LL, JEMD, LC: data abstraction and manuscript preparation; SK, KB, PvP, CW, JW-J, RT, RW, SRS, AM, TC, SS, BF, BE, ED, SO: data abstraction and multidisciplinary panel.

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