



National Steering Group for Specialist Children's Services

Paediatric Rheumatology Report

Introduction

1. The challenge of delivering high quality, accessible and sustainable services to children and young people in the context of the size and distribution of the Scottish population was recognised in the National Framework for Service Change (2005), “Building a Health Service Fit for the Future” (1).
2. The Minister for Health and Community Care established the Child Health Support group in 2000, to harness professional expertise in driving forward improvements in child health and the quality of child health services across Scotland.
3. HDL (2003) 43 outlined arrangements for a Scotland wide review of Children’s Specialist Services, which acknowledged the challenges faced by services with comparatively small numbers of patients and staff, coupled with demographic change and geographical considerations in respect of sustaining services. Paediatric Rheumatology was identified as one of the specialities for review.
4. An informal paediatric rheumatology network (Scottish Paediatric Rheumatology Group) comprising interested clinicians has evolved in Scotland over the last few years. This report reflects the views of clinicians working within that group; of a small multidisciplinary working party convened to produce this report and of parents attending recent network parent information days.

Background

1. The resourcing and development of Paediatric Rheumatology services in Scotland has lagged behind many other areas of the UK and many other paediatric subspecialties. The first and only full time NHS clinical post in the specialty was established as recently as 2005, several years behind other regions in the UK.
2. Until recently most children with rheumatic diseases in Scotland have been managed either by general paediatricians or adult rheumatologists with no specific training in paediatric rheumatology and there have been very few nurses or other Allied Health professionals with specific expertise in this area.
3. The Department of Health Musculoskeletal Framework (2) states that musculoskeletal conditions are the biggest cause of disability in children, accounting for failure to reach educational, social and physical milestones for many of those affected. Outcomes for children affected by Juvenile Idiopathic Arthritis have been poor with many of those affected reaching adulthood with significant disability and poor functional outcome including visual loss (3).
4. In recent years, the management approach to children with arthritis has changed significantly. The emphasis on early diagnosis, tight disease control and the availability of new drugs has led to a marked improvement in outcomes: disability is now largely preventable. All children with JIA should be screened for asymptomatic uveitis (4) to ensure that preventable visual loss is avoided. Children and young people with arthritis should now reasonably expect that their disease can be controlled and that they will enter adult life without significant disability or irreparable joint damage.
5. The improved outcomes achieved by current management practice dictate that if all children are to benefit they must have rapid access to assessment and advice from an expert specialty service. Delay in achieving disease control is one factor likely to be associated with adverse long term outcome (5). Although the situation has improved in recent years, this remains an aspiration rather than reality for many children in Scotland.
6. The British Society for Paediatric and Adolescent Rheumatology (BSPAR) Position Statement on Professionals Working in Paediatric Rheumatology (2007) (6) states that 'all Professionals working in the field of Paediatric Rheumatology need to demonstrate they have the required skills, knowledge and experience for this type of work' and that this applies to all members of the multidisciplinary team. It also states that children being managed out with tertiary paediatric rheumatology units should be managed as part of a Managed Clinical Network working closely with such a unit.
7. The establishment of a Senior Lecturer post in Paediatric Rheumatology in RHSC Glasgow 5 years ago provided the impetus to start the development of paediatric rheumatology services in Scotland and lay the foundation of the current informal clinical network in Scotland. The establishment of a full time NHS post in Paediatric Rheumatology shared between Edinburgh and Glasgow

2 years ago has enabled the service to develop further but clearly remains inadequate to provide access to specialist services for children throughout Scotland.

Incidence and prevalence of Rheumatology Disease

8. The scale of the problem and the service resources required to meet the needs of these children remains to be fully determined. Most children with rheumatic conditions are managed as outpatients and there is therefore no specialty specific data available from ISD codes.
9. The scope of the specialty and conditions included within its definition are clearly defined in the revised specialist definitions document produced for the Dept of Health by BSPAR, May 2007, (Appendix one).

Juvenile Idiopathic Arthritis

10. Juvenile Idiopathic Arthritis, the condition seen most frequently in paediatric rheumatology, is not an uncommon disorder.
11. The UK prevalence is estimated at 1:1,000 under 16 years with an incidence of approximately 1 in 10,000 (7). For the purposes of this report we have been able to obtain reasonably accurate figures for Grampian Region where we have identified 92 children with JIA (childhood population just under 100,000). Therefore although there is no accurate data currently available for Scotland, based on a childhood population of approximately 935,000, we would predict that there are over 900 children with JIA.
12. Clinicians currently working within the informal network for paediatric rheumatology have estimated that they are aware of around 500 affected children. Of these, many have received care in units with no particular specialty expertise. The estimated prevalence would suggest that there are several hundred more children in the country that we are unaware of. There are therefore many children in Scotland with arthritis who are failing to receive the benefit of appropriate specialist advice.
13. At a meeting of the Scottish Paediatric Rheumatology Group in 2006 it was agreed that the first task of a Managed Clinical Network (MCN), when established, would be to undertake detailed service mapping and establish the incidence and prevalence of rheumatic conditions in children in Scotland.

14. Despite the lack of published studies for comparison, it is clear that the outcome for children with arthritis in many parts of Scotland is suboptimal with an unacceptable number of children moving into adulthood with preventable poor outcomes, significant joint damage and visual loss. This is based on the clinical experience of a senior clinician who has worked elsewhere in the UK. ***'In two years working in Scotland I have seen more children with severe disability and visual loss from JIA than in 10 years working in Liverpool (a service for a comparable population size)'***, and on comparative data obtained from members of the British Society of Paediatric and Adolescent Rheumatology.

Other conditions

15. There are a relatively small number of children in Scotland (around 60 but possibly more) known to have more serious, complex connective tissue disorders, including SLE, juvenile dermatomyositis, scleroderma and systemic vasculitis. These children require management in, or in close collaboration with, a specialist unit.

16. There are small, but on a national basis significant, numbers of children with extremely rare but complex disorders that fall within the remit of paediatric rheumatology, e.g. CINCA, CRMO and Ehlers Danlos syndrome.

17. There are an unknown number of children with non-inflammatory musculoskeletal pathology, e.g. hypermobility and chronic musculoskeletal pain syndromes, who would benefit from access to specialist rheumatology multidisciplinary teams.

18. Improving outcomes means that accurate outcome data must be gathered. Standardised measures of outcome (8) are not being used in most units in Scotland seeing these children. It is hoped that an MCN would address this, facilitating the collection of standardised outcome measures in all units.

Current service provision

19. Current service provision is patchy. At present the only adequately resourced centre where children are guaranteed access to an appropriately trained team in a timely fashion is RHSC, Glasgow. There are significant inequities in service provision throughout the rest of the country.

Regional

Glasgow

20. There is now an established paediatric rheumatology team in RHSC, Glasgow comprising
- 1 WTE NHS Paediatric Rheumatologist (who also covers outreach to other units and provides services in Aberdeen and Edinburgh, 0.5 actually in Glasgow);
 - 1 Senior Lecturer in Paediatric Rheumatology (0.5 clinical; 0.5 academic);
 - 1 Paediatrician with an interest;
 - 1 WTE specialist nurse;
 - 1 WTE physiotherapist;
 - 0.5 WTE occupational therapist.

Edinburgh

21. Consultant Paediatric Rheumatology time in Edinburgh is provided by the 1 WTE NHS clinician under a Service Level Agreement (SLA) with Glasgow. This provides for 6 hours clinical work (MDT meeting plus 1 clinic) per week in Edinburgh plus 7 day telephone cover from the Yorkhill team. In addition Edinburgh has
- 0.5 WTE specialist nurse;
 - 0.4 WTE physiotherapist
 - 0.5 WTE occupational therapist.

22. The provision of 1 clinic per week is clearly inadequate for the needs of the population in Edinburgh. (Bristol which has a similar population base and sees very similar numbers of children with rheumatic disorders has two full time paediatric rheumatologists and one full time clinical nurse specialist.)

Aberdeen

23. Aberdeen has 1 specialist paediatric rheumatology clinic per month provided under a SLA with Glasgow, with support to the service provided locally by a general paediatrician with no previous training or experience.

Local

24. Two district general hospitals (Paisley and Wishaw) have Consultant Paediatric staff with some specific training and a declared interest in paediatric rheumatology, (Dr Jo Walsh and Dr Cath Lees).
25. In most other areas children are being cared for by general paediatricians and in some by adult rheumatologists. None have specific training and none are working with specialist paediatric rheumatology multidisciplinary teams. Some children are being seen in shared care clinics and other units working within

the network have requested shared care clinics but resources are insufficient to provide this.

Specialist Nursing Posts

26. There are 1.5 specialist nurses in Scotland (based in Glasgow and Edinburgh, as described above) who support local services but have insufficient time to provide backup and support for other units.
27. One of the major roles for nurses within the specialty is the training of families to give parenteral therapies at home; the supervision of children receiving these treatments; providing telephone advice to families administering such treatments in a home environment and ensuring that the drugs are being appropriately monitored. The lack of CNS to undertake this role is a significant clinical risk within the speciality. Parents (see Appendix three) describe being unable to access support if they have concerns re their child's drug therapy and monitoring in some areas is less than ideal.
28. Appendix two summarises the current workforce in the speciality.

Proposed model of care

29. The preferred model of care is to enable local teams to deliver evidence based treatment, with support from a centrally based National team with specific Paediatric Rheumatology expertise and training. All local teams have expressed a willingness to work as part of an MCN with shared care clinics being an integral part of service delivery.
30. There are particular issues in trying to develop locally delivered care in paediatric rheumatology. Unlike most other specialties, paediatricians very rarely receive even basic experience in the specialty during their general training. Even if resources permitted, it would not be possible to appoint locally based paediatricians with an interest in paediatric rheumatology, as there are none trained
31. In order to provide optimal locally based care, there is a need to identify a network of Paediatricians who are willing to develop a 'special interest' and provide them with appropriate training and support. Dedicated time in the clinicians' job plan will also be required to enable them to fulfil this role.
32. The development of local teams to provide a high standard of care is therefore dependent on the central 'national' team being adequately resourced to provide the necessary education, training and support. At present this is not the case. The development of a formal MCN would hopefully provide the necessary administrative support to aid in the development and organisation of this, but this needs to be backed up by an

expert clinical team with time to deliver the required training. This 'national' team should comprise of a lead clinician, lead nurse and lead physiotherapist to address the clearly identified training and service needs around the country.

33. Each local service requires to ensure that the child / young person should have access to a multidisciplinary team comprising Doctor, Nurse, Physiotherapist, Ophthalmologist and Occupational Therapist.
34. Each local team will require adequate support from pharmacy, radiology and clinical psychology to address the needs of these children
35. Each local service will work with a local adult rheumatologist to develop transition services for children in their locality.
36. There is also a need to work with primary care to ensure that children with rheumatic disorders are recognised and referred in a timely fashion to an appropriate service. It is hope that primary care involvement in the MCN can facilitate this.

Parent views (see Appendix three for more detail)

37. While most support locally based care, parents clearly identified the need for more specialist medical expertise. Some would travel for this if it cannot be provided locally, however, many parents would find this extremely difficult. Many had noted clear benefits to their child from a change in management when seen in a specialist clinic, while others expressed frustration that they had not had this opportunity, or that it had come very late in their child's illness when irreversible joint damage had already occurred. Those parents not attending Glasgow or Edinburgh were frustrated by their lack of access to the support that a specialist nurse would give.
38. The overall aim is to develop a model of local teams working with a national team of experts within an MCN. This should ensure that children have appropriate and equitable access to the best treatment available for their condition.

Standards of Care and Guidelines

39. Children with rheumatological conditions require access to specialist care provided as locally as possible and in an age appropriate environment. Numerous documents highlight the need for children to be managed by paediatric specialists in a paediatric environment.

***'It seems so obvious it hardly needs to be said: just as children differ from adults in terms of their physical, psychological, intellectual and emotional development, so they differ in their healthcare needs. They experience and see the world differently. Children are in a constant state of growth and development which creates particular needs and demands which are of a different order from those affecting adult patients. Their relative physical and emotional immaturity in comparison with adults, has implications both for the treatment they receive and the physical environment in which they are cared for.'*(9)**

40. Children therefore need access to care that is appropriate to their age but also meets the needs of their condition. All children should have access to a multidisciplinary team with appropriate specialist expertise. The BSPAR Position Statement on Professionals working in Paediatric Rheumatology (2007) (6) clearly documents that members of these teams must have the appropriate skills, knowledge and training to work in this area.
41. Expert opinion internationally agrees that changes in approach to treatment and available drug therapies over the last decade have resulted in significantly improved outcomes for patients in centres adopting current best practice. Data remains anecdotal: changes in treatment are relatively recent and results of outcome studies are awaited.
42. Children must be referred to an appropriate service in a timely fashion. Delay in the diagnosis and treatment of inflammatory arthritis is associated with a worse outcome (5) Standards of Care produced by ARMA (Arthritis and Musculoskeletal Alliance) (10) recommend that all children suspected of having an inflammatory arthritis should be seen in a specialist clinic within 4 weeks.
43. Children at the complex end of the spectrum require management to be co-ordinated from a tertiary centre with access to specialist expertise from other services, eg cardiology, nephrology, immunology.
44. Silent or asymptomatic uveitis associated with this condition is a preventable cause of visual loss in children. Guidelines produced jointly by BSPAR and the Royal College of Ophthalmology (4) define recommended intervals for Ophthalmological screening for children with JIA. Asymptomatic patients should be screened as a clinical priority, no later than 6 weeks after referral. Symptomatic children should be seen within one week of referral. Recommendations for screening of high risk patients are for 2 monthly screening for 6 months and then 4 monthly until the age of 11 years. In many units these recommendation are not being followed and many children with JIA receive no or inadequate eye screening.
45. There is now an emerging evidence base for the use of drugs in juvenile arthritis (JIA). Methotrexate had been shown to be effective for disease control and should to be offered as soon as possible after diagnosis to all children with polyarticular JIA. The supervision and monitoring of methotrexate(11) is best undertaken by a clinical nurse specialist. In many

children, this is better tolerated and more effective when given subcutaneously. All children requiring parenteral therapies should have the option of this being delivered by the family at home with the support of a specialist nurse (12).

46. Intra-articular steroids injections are an effective method of obtaining rapid disease control and symptom relief. All children with JIA should have rapid access to joint injections in an appropriate environment as necessary. For very young children this requires access to general anaesthesia and an appropriately trained person to undertake the injection. Older children should have the option of using entonox to facilitate the injection (13) where possible in the outpatient department to minimise return appointments.
47. Many units do not use triamcinolone hexacetonide (TH) for joint injections in children despite evidence of its superior efficacy in paediatric practice(14). The use of TH minimises the need for repeat injections (with in many cases repeat general anaesthesia and daycase admissions). All units offering joint injections to children should use TH rather than cheaper but less effective alternatives.
48. NICE guidelines(15) use clearly defined criteria for access to etanercept one of the anti-TNF agents. This drug is effective for many children who have failed to respond to other therapies and significantly improves outcomes for those with more severe disease. Despite this there is significant variation across Scotland in the provision of etanercept to children who meet criteria.
49. NICE guidelines state that etanercept should only be prescribed in conjunction with a specialist unit, that it must be supervised by a specialist nurse and that data on children on these drugs should be entered onto the BSPAR National Biologics register. This guidance is not being adhered to in much of Scotland due to lack of adequate resourcing.
50. The role of the specialist nurse in managing and monitoring these therapies is clearly defined in the appropriate RCN guidance (16).

Resource issues

National

The establishment of a National Managed Clinical Network for Paediatric Rheumatology will facilitate the delivery of the service model outlined in this document.

- The development and submission of the application to NSD for the MCN is currently being delayed by the acute resource shortage affecting the specialty.

- There is an urgent need for Scotland-wide guidelines: e.g. ophthalmology screening; drug administration and monitoring. The establishment of the MCN will provide a mechanism for the development and dissemination of such guidelines.

Medical resource:

- A sustainable Scotland wide service requires 3 WTE NHS Consultants in the speciality plus the current Senior Lecturer post. The need for additional Consultant specialty expertise is immediate.
 - The siting of these posts can be explored but the current model with a core team based in Glasgow and Edinburgh providing outreach to other areas would require two clinical posts based in Glasgow and one in Edinburgh. An alternative model would be to have all three clinical posts based in Glasgow with outreach from there to all other centres.
 - There is an immediate need for a further fully trained WTE Consultant if service delivery is to be sustained and progress maintained.

Nursing and AHP:

- 1 Nurse Consultant in Paediatric Rheumatology to support, develop and work with locally based teams.
- 1 Physiotherapy Consultant / advanced practitioner in Paediatric Rheumatology to support, develop and work with local teams.
 - This is in addition to further physiotherapy time required to sustain the local service.
 - This model of development has been discussed with and is being proposed with the agreement of the Heads of Paediatric Physiotherapy services in RHSC Glasgow and Edinburgh and Royal Aberdeen Children's Hospital.

Regional / local

Medical

- Identified Paediatrician with an interest in each District General Hospital.
- Identified Ophthalmologist in each DGH to take responsibility for the ophthalmological screening of children with JIA.

Nursing

- Adequate Specialist Nurse time in the Children's Hospitals: establish a part time post in Aberdeen and increase Edinburgh post to full time, (Total 1 WTE).
- Named nurse for paediatric rheumatology in each DGH.

Allied Health Professionals

- Dedicated Physiotherapy time in every local area.
- Access to Paediatric Occupational Therapist.

Other services required in Units wishing to provide a local service

- Pharmacy – new drug regimes are placing significant demands on pharmacy services, and District General Hospital Pharmacies are not always equipped with the resource, knowledge and expertise required.
- Radiology
- Psychology
- Social work
- Play therapist.

Interdependencies with other services

IT support.

51. Adequate IT support is required to facilitate the proposed model of care. Children who are being managed locally, but with occasional visits to or from the specialist team, may end up with partial documentation of their illness in each of several sets of notes and with none of the health care professionals responsible for their care having access to the complete record. With the use of complex cytotoxic and immunosuppressive drug regimes for these children this is a significant area of clinical risk and must be addressed.
52. For the national team working across many centres the multiplicity of case notes and electronic systems that are required to work in each centre is impossible to manage.
53. The procurement and implementation of a single electronic system to be used throughout Scotland as recommended in 'Building a Health service Fit for the Future' (1) is essential if small specialties are going to be able to offer effective care on a Scotland-wide basis.
54. Specialist Paediatric Rheumatology services require close liaison with many other specialties. Many rheumatological disorders are multisystem conditions and require assessment by and liaison with many different specialists. These include:
 - Ophthalmology, Endocrinology, Immunology, Orthopaedic surgery, Nephrology, Cardiology, Dermatology, Haematology; CAMHS, Maxillofacial/dentistry, metabolic bone disease, Genetics, Gastroenterology and transition to Adult Rheumatology.
 - Paediatric anaesthesia and theatre space for intra-articular steroid injections in the young child. Nursing staff trained to administer entonox to facilitate injections in older children.

- Access to PICU care for seriously unwell children with multisystem disorders.
- Children with rheumatic disorders require access to a Specialist Allied Health Professional Team, including Nursing, Physiotherapy, Occupational Therapy, Podiatry, Psychology, Orthotics and Dietetics for assessment, education, treatment and rehabilitation programmes.
- Paediatric Radiologist with expertise in musculoskeletal radiology.
- Specialist liaison with Education and Social Services to address needs of specific chronic illness.
- Specialist emotional and spiritual support to meet the needs of family and child with chronic illness.
- Expertise in assessment of neglect, emotional abuse and child protection for children with chronic illness and disability.

Risks

55. The management approach to rheumatic disease has changed significantly over the last few years.

Prompt referral at diagnosis to a specialist, and the establishment of correct and appropriate treatment with regular follow up and monitoring has significantly improved the lives of children. Long term disability and visual loss is preventable in the majority with a marked reduction in the need for joint replacement surgery in adult life. This will only be achieved when all children are able to access appropriate specialist care.

56. A key issue for the service in Scotland is a shortfall in 'specialist expertise' at Consultant, Nurse and Physiotherapy level.

57. Currently the specialist Consultant time required to support regional / local care and its development is not available. The result of this is some Health Board areas are not treating children optimally. This results in long term health / treatment issues with subsequent 'cost' to the child's quality of life, and the financial costs of poorly controlled disease in terms of increased ongoing medication costs and surgery including joint replacement.

58. The Edinburgh Rheumatology service is currently unsustainable. Consultant input is provided via a joint appointment between Edinburgh and Glasgow. This was funded to provide a clinic 1 day / week in Edinburgh, but has been informally increased to 2 due to demand. This is still inadequate for the demands of the Edinburgh service and is now taking Consultant time away from Network development and the Glasgow service. It will revert to 1 day / week at the end of December 07 leaving the Edinburgh service unsustainable.

Nursing and Physiotherapy support in Edinburgh are also inadequate to sustain the service.

59. The current provision of 1.5 WTE Specialist Nurse posts in Glasgow and Edinburgh mean that care and support to children and families, but also support and education for Nurses at a local level is not achievable. The 2 Nurses are working at capacity locally providing education, training and support to families administering new therapies. Locally based hospital or community based children's nurses could be trained to provide some of this and to support families locally, but this would require an investment of time and expertise from the national team.
60. A Nurse consultant would be ideally placed to deliver such training on a national level.
61. An additional 1WTE specialist nurse is needed to increase the level of support in Edinburgh and provide some much needed support to families in Aberdeen.
62. Physiotherapy provision across the country is patchy. Many children are treated by their locally based community children's physiotherapists who have no specific training in the care of children with JIA. Children with rheumatic disease require access to Physiotherapists who are able to regularly update their knowledge and receive appropriate support to ensure optimal care. A national lead post would be ideally placed to provide the education and support required. The establishment of such a post is supported by the heads of physiotherapy in RHSC Glasgow and Edinburgh, and Aberdeen Children's Hospitals.

The care of, and consequently the outcome for, children with arthritis remains suboptimal in many areas of Scotland. Over the last few years an informal network of interested clinicians has developed but the resource to develop this further into a network of expert local teams is lacking. This must be addressed urgently to prevent further children developing preventable disability.

Recommendations:

Recommended Action	Timescale
Appoint 1 WTE Paediatric Rheumatologist Provide a service in Edinburgh Outreach clinics: Fife Tayside	Immediate
Establish Managed Service / Clinical Network, prepare and submit bid to NSAG	2008
Appoint 0.5 WTE Specialist Nurse Grampian To establish an urgently required resource for children and families Appoint WTE Consultant Nurse	2008 2009
Appoint Consultant/advanced practitioner Physiotherapist	2009
Work via MCN to ensure implementation of ophthalmology screening guidelines for all children with arthritis via named ophthalmologist for each area	2009
Regional Planning teams to scope / establish robust data to support local service development and work with MCN in its delivery. .	2009
To facilitate planned development of a parent network	2009

EXTRACT FROM: SPECIALIST SERVICES NATIONAL DEFINITION SET: PAEDIATRIC RHEUMATOLOGY

DRAFT DOCUMENT PREPARED BY BSPAR FOR DOH MAY 2007

Rheumatic diseases are an important cause of disability in childhood. Proper diagnosis and early aggressive intervention can minimize both short and long term morbidity of these conditions. Without proper therapy, acute rheumatic fever, systemic lupus erythematosus, dermatomyositis, progressive systemic sclerosis, severe systemic arthritis and many forms of vasculitis can be fatal. Other conditions such as juvenile idiopathic arthritis and spondyloarthropathies which do not acutely threaten life, can be associated with lifetime disability. However, the diagnosis of rheumatological disorders can be difficult and there is often delay in appropriate referral which has been shown to worsen the outcome in inflammatory arthritides and probably in inflammatory disorders in general.

There are important age related impacts of the diseases on the developing immune, neurological and musculoskeletal systems. These chronic diseases have profound psychosocial effects on patients and their families. Many of the diseases are very rare. Even the more common juvenile idiopathic arthritis (with its 7 subtypes) has an incidence of only 1/10,000 and a prevalence of 1:1000. Other conditions such as juvenile dermatomyositis are extremely rare. In rare illnesses the care must be by specialists in order for the clinical teams to have the necessary experience and expertise to diagnose and treat safely.

The first goal of treatment is to make the correct diagnosis and this itself is often a specialist task in this specialty. In paediatric rheumatic diseases, findings on physical examination often take precedence over laboratory findings in the establishment of a diagnosis and a treatment plan. The importance of a skilled and appropriately trained examiner cannot be over emphasized. It is therefore difficult to make a clear generalist/ specialist split for a particular disease prior to specialist referral. The next aims are to gain rapid control of disease activity, preserve normal physical, social and emotional growth and development, minimize chronic disability and deformity, and achieve and maintain remission of disease.

Trained paediatric rheumatologist are highly skilled in:

- 1) evaluation of difficult examination findings,
- 2) differential diagnosis in children and adolescents;
- 3) efficient use of diagnostic interventions;
- 3) selecting the most appropriate therapy (including other consultative services)
- 4) monitoring long term therapy for effectiveness and side effects unique to children and adolescents;
- 5) achieving favourable outcomes in terms of control of rheumatologic diseases and prevention of disability;
- 6) coordination of care for children and adolescents with multi-system diseases; and
- 7) dealing with chronically ill children, adolescents and their families.

Links with other specialist services definitions set

Many disorders of childhood may have a rheumatological component and many rheumatological disorders are multi-system in their nature. Very close collaboration and often joint clinics are needed with the following paediatric specialties:

- a) Paediatric orthopaedics,
- b) Paediatric nephrology and access to renal biopsy service,
- c) Paediatric ophthalmology,
- d) Paediatric cardiology with access to assessment of pulmonary hypertension in collaboration with supra-regional pulmonary hypertension centres,
- e) Paediatric respiratory medicine and access to comprehensive lung function testing

- f) Specialised child psychology/psychiatry,
- g) Paediatric dentistry, orthodontist and maxillo-facial surgery,
- h) Paediatric dermatology with access to dermatology pathology services
- i) Paediatric gastroenterology and nutrition
- j) Paediatric immunology
- k) Paediatric theatre, anaesthetics
- l) Paediatric intensive care
- m) Paediatric endocrinology
- n) Specialist paediatric radiology with expertise in musculo-skeletal ultrasound, MRI and CT scanning, videofluoroscopy
- o) Specialist speech and language service for swallowing disorders in scleroderma
- p) Genetics service,
- q) Links with skeletal dysplasia network and metabolic bone disease specialist (this is an area that needs to develop in paediatrics in association with paediatric rheumatology
- r) Paediatric oncology/ haematology team for diagnostic bone marrows and joint management of patients
- s) Specialist pathology services for synovial fluid cytology, and histological diagnosis and many organs including muscle, renal, gut ,mouth, bone, meninges and rarely brain.
- t) Paediatric Ophthalmology for complex uveitis cases.

Specialised Paediatric Rheumatology Services are needed for the following situations:

- Patients with unclear diagnoses or complex multi-system presentations
- Prolonged fever, rash ,arthritis, weakness, weight loss, anaemia raised inflammatory markers or generalised malaise, fatigue or anorexia
- Rashes in the presence of any of the above, or vasculitic or typical rheumatological rash e.g. of SLE or systemic onset JIA
- Chronic medically unexplained pain or loss of function, or complex chronic fatigue
- Undefined autoimmune disease e.g. Complex autoimmune thrombocytopenia
- Patients with Connective Tissue Diseases including Systemic lupus Erythematosus, Juvenile dermatomyositis, Mixed connective tissue disease , Scleroderma – systemic and localized/ linear morphoea, Sjögren’s Syndrome
- Patients needing immunosuppressive, cytotoxic or biologic therapies
- Patients with familial periodic fever syndromes
- Reactive (post infectious) arthritis
- Diagnostic evaluation and long-term management of: Juvenile idiopathic arthritis. JIA subtypes oligo articular (persistent or recurrent)
- Chronic vasculitis
 - Polyarteritis nodosa
 - Atypical Kawasaki disease
 - Atypical Henoch Schonlein purpura
 - Wegner’s granulomatosis
 - Behcet’s syndrome
 - Takayasu’s arteritis
 - hypocomplementemic vasculitis or hypersensitivity vasculitis
 - Cerebral vasculitis
 - Post-infectious vasculitis
- Anti-phospholipid syndrome
- Acute rheumatic fever
- Sarcoidosis
- Lyme disease with arthritis
- Chronic recurrent multifocal osteomyelitis
- Neonatal onset multisystem inflammatory disease /CINCA
- Post-infectious arthritis
- Relapsing polychondritis
- Iritis
- Osteoporosis
- Raynaud’s phenomenon

- Osteochondroses
- Genetic syndromes associated with stiff joints or severe hypermobility e.g. Ehlers Danlos and Marfan's syndrome
- Joint disease associated with other medical diagnoses e.g. inflammatory bowel disease, cystic fibrosis, complex cyanotic heart disease, Down's syndrome, immunodeficiency, neoplasm, infectious disease, endocrine disorders, genetic and metabolic diseases, post-transplantation, and arthritis associated with birth defects.
- Difficult Child Protection cases presenting with rheumatological features
- Major pain syndromes
 - Erythromelalgia
 - Fibromyalgia
 - Reflex sympathetic dystrophy and complex regional pain syndromes
 - Cold induced injury
 - Over use syndromes

APPENDIX TWO			
Hospital	Medical	Specialist Nurse	Physiotherapy
Aberdeen Children's Hospital	Dr W Houlsby (general paediatrician) Dr J Davidson (paediatric rheumatologist, 1 clinic per month) Dr N Rennie (adult rheumatologist, 1 clinic per month)	Support from staff in medical day unit	Alison Aiken (not specific paed rheum post but sees all local paed rheum patients)
Dundee	Dr P Fowlie (general paediatrician) Dr K Morley (adult rheumatologist, 1 clinic alternate months)		
Fife	Dr J Gibson (adult rheumatologist with special interest)		
Edinburgh	Dr J Davidson (paediatric rheumatologist, 1 day per week)	Imogen Kelly 0.5 WTE	Jenny Graham (post shared between several specialities; approx 0.3 paed rheum)
Stirling/Falkirk	Dr M Brzezki (Adult Rheumatologist)		
Lanarkshire	Dr C Lees (paediatrician with an interest) Dr E Murphy (adult rheumatologist with an interest) Clinic with J Davidson every 3 months		
GGC	Dr J Davidson (paediatric rheumatologist) Dr P Galea (paediatrician with an interest) Dr J Gardner-Medwin (senior lecturer) Dr J Walsh (paediatrician with an interest)	Vicki Price 1.0 WTE	Caroline Grant and Audrey Ritchie (1WTE: also contribute to general Musculo Skeletal paed service)

	interest; Paisley)		
Borders	Dr J Stephen (general paediatrician		
Dumfries	Dr Robert Simpson (General paediatrician) Clinic with Dr J Davidson 2x per year		

Rheumatology Parents day – Aberdeen Sat September 15th

Feedback from parents

Thirty four parents attended the event. The majority were from Grampian, with parents also from Edinburgh, Fife, and 1 from Shetland. The morning was a series of information sessions about JIA, and 2 parents shared their experiences. During the course of the afternoon, 5 workshops ran, with the parents split into smaller groups attending all of them.

During the course of the afternoon, the 5 groups had an opportunity to reflect on the following questions:

- What is good / what do you value about the service you receive now?
- What improvements would you like to see in the future?

The following is a summary of views expressed.

It quickly became apparent that the parents from Edinburgh and Fife (who were accessing the Edinburgh service), valued access to a ‘team’ of people all dedicated to Rheumatology. This was described by the parents clearly as containing the following people:

Consultant Rheumatologist
Specialist Nurse
Physiotherapist
Social Worker (7 hrs / week)

They described a service which had a coordinated and consistent approach to their needs, and clearly articulated the value of the Specialist Nurse who they could phone throughout the week for advice (currently a part time post).

Grampian parents had a different experience to share. They described a service that did not appear to have a team approach, but individuals within it were really appreciated. The Physiotherapist, Nurses on the day unit, Dr Davidson, (Consultant Paediatric Rheumatologist) Dr Houlsby (Paediatrician) and Dr Rennie (Adult Rheumatologist). It quickly became evident that the parents who saw Dr Davidson were a small minority (Wednesday clinics), and raised the question from others about who decided which children saw Dr Davidson. Some parents had been referred to Yorkhill to see her, and were then followed up locally. 1 parent had referred themselves to Yorkhill after a long protracted period when the diagnosis was not known

Summary of points raised in response to the questions

Information

Parents valued the information provided though the ARC website (Arthritis Research Campaign), but felt that a locally produced support pack for schools would be beneficial as the ARC pack is very detailed. It was felt important that schools had timely, appropriate and up to date information.

A parent held record / careplan was discussed as an idea with one group, and was warmly received to save having to tell everyone time and time again the same information.

The time around diagnosis is a blur and written easy to follow information handed out by the hospital would be really beneficial.

Access to continuing support

This came up in nearly every group. There were understandable anxieties that local primary care teams were not equipped with the knowledge about the current medication and management of the disease. Parents described various situations where they had had a query about medication because their child had been unwell, or had vomited after taking it etc. All who had experienced trying to access information in Grampian described problems unless the Consultant was available, most appeared to have access to his mobile or e mail, but this was not helpful when he is on holiday! One parent was routed to a cancer doctor as the query was in relation to Methotrexate.

This need would be addressed by the appointment of a nurse specialist.

Drugs

The new drug therapies available for this disease are not generally known about, 'disease modifying anti rheumatic drugs' e.g. Methotrexate, and 'Biologics' eg Enbrel. These are usually administered by the parents at home, the former is given orally or by sub cutaneous injection on a weekly basis, the latter twice weekly as a s/c injection. One parent described being given the prescription for Enbrel, but it took 4 months for the script to be dispensed. The parent was not sure what the problem was, and the Consultant was not aware there had been a problem until the next clinic visit, 6 months after the original prescription had been handed out in clinic. This was felt by the parent to be due to a lack of follow up and demonstrated a break down in communication

Others talked about the difficulties taking in information about the drugs being prescribed 'there is never enough time', and no one to follow up a few days after the clinic appointment to check everything is ok. A phone call would be valued to allow parents to check out the information they have been given and clarify understanding.

Treatment in general

Those who had experienced moving between different hospitals described treatments being done differently. Consistency of approach appeared to be different in respect of procedures, joint injections for example.

Attitudes to children in pain appeared to be markedly different – ‘yorkhill will not accept a child in pain’ was a clear message from a mother who had watched her child suffer for months. (This may be due to the presence of a pain team and heightened professional awareness.)

There was a strong feeling that specialist input was required even if there is an identified Paediatrician who has developed an interest. The Grampian parents asked the Edinburgh parents to describe what a Specialist Nurse could do.

The Lady Hoare trust Social Worker in Grampian used to be a real support for these families, and provided a link in the community between hospital visits. The families described warmly how she was there to listen, help them source the right support, passed information into the school, and generally be available to support them.

Local GP carrying out blood tests really appreciated to save unnecessary trips to hospital.

What would make the service better?

- A team approach in Grampian please with some specialist nursing input.
- Annual day for parents to meet together and share.
- More of ‘Dr Davidson’.
- A way of putting the children in touch with each other via the web or organised days. (Parents described their children not knowing anyone else with the disease).
- A proper medical service for the number of children.
- Parent held record to allow a feeling of control.
- 24 hr access to an advice line for information, particularly in relation to symptom management and drugs.
- Appointments on the same day to save trips to hospital.
- A central data base in Scotland which parents could access and update on a regular basis to help with research, the parents identified they want to do something to help.

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