One Hundred and Seventy Cases of Childhood Onset Rheumatological Disease in Singapore

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Abstract

One hundred and seventy patients with rheumatological disease diagnosed before their 16th birthday and still on follow up were studied retrospectively. They were seen within the last 3 years at KK Women's and Children's Hospital, Tan Tock Seng Hospital, National Skin Centre or Singapore General Hospital. Of these, 89 were still less than 16 years old at the time of study.

The majority had systemic lupus erythematosus (51.8%). Many were on long-term follow-up for persistent disease, including renal manifestations (47.7%), neurological manifestations (26.1%) and haemolytic anaemia (15.9%). Photosensitivity and malar rash were more common than in Western studies while arthritis was less common. Anti-phospholipid antibodies were found in children complicated by myocardial infarction, pulmonary hypertension, Raynaud's phenomenon, cerebral and gut lupus.

Children with juvenile chronic arthritis comprised 28.8% and juvenile dermatomyositis 10%. The male predominance and lack of uveitis in children with pauciarticular JCA were striking. Rarer conditions included polyarteritis nodosa, scleroderma, rheumatic fever with arthritis, polychondritis and Behcet's disease.

Many diseases may first present with a rheumatological complaint. This review of features of local children highlights similarities and differences with Western data. It also provides information for planning long-term care, multidisciplinary clinics, group physiotherapy sessions, educational programmes and support groups.

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Introduction

Children with rheumatological disease form a small but important proportion of the ill paediatric population. Most of the published data available are based on studies on Caucasian, Black or Hispanic children with relatively little information on Asian children including Chinese and Malays.

The aims of this study were: 1) to collect data on children with rheumatological disease locally—demographic, clinical, complications and treatment response and to compare this with published data on children or local Asian adults, and 2) to better understand the scope of the disease locally and to plan for more complete and appropriate management.

Materials and Method

A paediatric rheumatology registry was set up in March 1997. Patients diagnosed to have rheumatological conditions with age of onset before their 16th birthday were entered into the registry. They had been admitted or seen as outpatients within the last 3 years at KK Women's and Children's Hospital (KKH), Tan Tock Seng Hospital (TTSH), National Skin Centre (NSC) and Singapore General Hospital (SGH) by paediatricians, rheumatologists or dermatologists.

Patients with the following diagnoses were included: juvenile chronic arthritis (JCA) including spondyloarthropathies (SPA), systemic lupus erythematosus (SLE), juvenile dermatomyositis (JDM), overlap syndromes [or mixed connective tissue disease (MCTD)], scleroderma, polyarteritis nodosa (PAN), polychondritis, other vasculitides, rheumatic fever, chronic recurrent multifocal osteomyelitis (CRMO), Behcet's disease, reflex sympathetic dystrophy (RSD) and hypermobility.

Reactive arthritis and Henoch-Schonlein purpura (HSP) were not analysed as most cases were uncomplicated and not followed up for long. Also, because the most significant complication of patients with HSP is

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usually renal, such patients are followed up by the nephrology service rather than the rheumatology service. Kawasaki disease was not entered into the rheumatology registry as its main complication is cardiovascular and such patients are followed up by cardiology or general paediatrics. Patients with "orthopaedic" diagnoses such as Perthe's disease, Slipped Capital Femoral Epiphyses (SCFE) and septic arthritis were also excluded. Patients presenting with arthritis but found to have leukaemia or other malignancies were referred to the relevant subspecialists. However, patients who may benefit from programmes offered by paediatric rheumatology such as physiotherapy and hydrotherapy classes were entered into the registry, e.g. RSD and hypermobility.

Criteria for the rheumatological diseases studied are as follows:

Age of onset before the 16th birthday

a) Systemic lupus erythematosus (any 4 of the following 11 features):
   - malar rash
   - discoid rash
   - photosensitive skin eruption
   - oral / mucosal ulcers
   - non-erosive arthralgia / arthritis
   - serositis
   - renal involvement of proteinuria >0.5g / day or cellular casts
   - neurological involvement e.g. psychosis, seizures, transverse myelitis, peripheral neuropathy, cerebral arteritis
   - cytopenia such as haemolytic anaemia, thrombocytopenia or leukopenia
   - positive anti-nuclear antibody by immunofluoresence
   - other positive serology (antibody to double-stranded DNA, antibody to Smith antigen, antiphospholipid antibody, false positive VDRL)

b) Juvenile chronic arthritis
   Arthritis for at least 3 months, with no other known cause
   - *Still's Disease:* characteristic high spiking fever with arthritis
   - *Polyarticular disease:* >4 inflamed joints by 6 months after onset of illness
   - *Pauciarticular disease:* ≤4 inflamed joints by 6 months after onset of illness
   - *Spondyloarthropathy:* arthritis involving axial skeleton (such as sacroiliac joints, spine) as well as peripheral joints especially if with enthesopathy, tenosynovitis, plantar fascitis, HLA B 27 positivity and iritis

c) Juvenile dermatomyositis
   Characteristic skin inflammation (heliotrope rash, Gottron papules or vasculitis) with myositis or myopathy (symmetrical proximal weakness and raised muscle enzymes or electromyograph showing myopathy and denervation or muscle biopsy showing necrosis and inflammation) as well as absence of laboratory markers of SLE or MCTD

d) Mixed connective tissue disease
   Arthritis with features of dermatomyositis, scleroderma and SLE with very high RNP antibody titre

e) Chronic recurrent multifocal osteomyelitis
   Recurrent inflammatory multifocal lytic bony lesions with no other known cause

f) Scleroderma
   Tightening and thickening of skin and subcutaneous tissue either localised or systemic

g) Polyarteritis nodosa
   Necrotising vasculitis of small and medium sized muscular arteries with formation of nodules in the absence of obvious allergy, significant eosinophilia or pulmonary lesions

h) Polychondritis
   Inflammation of cartilage with widespread systemic features including vasculitis and arthritis with no other known cause

i) Behcet's disease
   Recurrent genital and oral ulceration with relapsing iritis and or conjunctivitis

j) Rheumatic fever
   Two major criteria (carditis, migratory polyarthritis, chorea, erythema marginatum, subcutaneous nodules) or 1 major and 2 minor criteria (fever, arthralgia, prolonged PR interval, elevated erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP), past history of rheumatic fever) with evidence of recent streptococcal examination (rising or significant anti-streptolysin O titre, positive pharyngeal cultures)

k) Benign symptomatic hypermobility
   Hyperextensibility of joints associated with musculoskeletal pain but no obvious arthritis or underlying connective tissue diseases such as Ehlers Danlos or Marfan's syndrome

l) Reflex sympathetic dystrophy
   Sudden bizarre immobile and painful posturing of limb with colour or trophic changes and no evidence of arthritis or other known pathology

Patients with the above rheumatological diagnoses and currently on follow up had their demographic, clinical, key investigative and treatment data collected from medical records from 1 March to 31 July 1997 and entered into a standard data collection sheet. Data were entered into the paediatric rheumatology registry. Data collected from 1 March to 31 July 1997 were then analysed for this report. However, data entry into the regis-

try continues and this will be useful for future analysis.

One drawback of this study is that the data only captured patients who were alive and still on follow up at the time of study. Patients who recovered and were not followed up, patients who had died and patients who had defaulted or left the country could not be analysed. Certain groups of patients in which the majority are not followed up by rheumatologists e.g. Kawasaki disease, Henoch-Schonlein purpura and reactive arthritis were not analysed. Current data of surviving patients are important as it provides information for the planning of health services for patients. Follow up of all current patients will provide data on disease patterns, mortality, morbidity and dropout rates. This is a cross-sectional assessment, but prolonged follow up will yield greater information in a longitudinal study.

Results

Of the 170 patients entered into the registry with disease onset before their 16th birthday, 89 were still below 16 years of age (Table I). The majority had systemic lupus erythematosus (51.8%). Juvenile chronic arthritis (including spondyloarthropathy) comprised 28.8% while juvenile dermatomyositis comprised 10%. In the group with juvenile spondyloarthropathy a proportion continued to have active disease or developed ankylosing spondylitis (AS) with increasing age. Rarer conditions include polyarteritis nodosa, scleroderma, rheumatic fever with arthritis, polychondritis and Behcet's disease.

The majority of the rheumatology patients were female (ratio 2.2:1) with a female predominance for SLE, JDM, overlap syndrome and polyarticular onset JCA. There was a male predominance for SPA, AS and pauciarticular onset JCA, and a similar sex distribution for Still's disease. The majority of patients were Chinese with a smaller proportion of Malays, Indians and other races reflecting our population's composition. Demographic data for Singapore's population are shown in Table II for comparison.[1,2]

Systemic Lupus Erythematosus

SLE formed the largest group of patients in the paediatric registry still under follow up. The oldest patient with paediatric onset disease was 44 years old at the time of study and has been followed up for more than 28 years (she was 15 years old at diagnosis). Of the 88 patients, 30 (34%) were still under the age of 16 years, and 65 (74%) under the age of 21 years. The majority presented above

TABLE I: TYPES OF PAEDIATRIC RHEUMATIC DISEASES SEEN IN THIS STUDY

	Total	Female	Male	Age <16 years
SLE	88(51.8%)	74	14	30
JDM	17(10.0%)	14	3	13
Overlap	4 (2.4%)	4	0	2
JCA/SPA	49(28.8%)			
Still's	6	4	2	4
Pauciarticular JCA	13	5	8	12
Polyarticular JCA	10	6	4	5
SPA (including Psoriatic)	18	6	12	13
AS	2	0	2	0
Behcet's	1	0	1	1
PAN	1	0	1	0
RSD	1	0	1	1
Rheumatic fever	1	0	1	1
CRMO	1	0	1	1
Scleroderma	1	1	0	1
Hypermobility	5	2	3	4
Polychondritis	1	1	0	1
Total	170	117	53	89

AS: ankylosing spondylitis; CRMO: chronic recurrent multifocal osteomyelitis; JCA: juvenile chronic arthritis;
JDM: juvenile dermatomyositis; RSD: reflex sympathetic dystrophy;
PAN: polyarteritis nodosa; SLE: systemic lupus erythematosus;
SPA: spondyloarthropathy;

TABLE II: AGE OF ONSET, SEX RATIO AND RACIAL DISTRIBUTION OF PAEDIATRIC LUPUS COMPARED TO LOCAL DEMOGRAPHIC DATA

Lupus onset age (y)	Number (%)	Female	Male	F:M
<5	2 (2.3%)	2	0	
5 to 10	18 (20.5%)	14	5	2.8 :1
10 to 16	68 (77.3%)	58	9	6.4 :1
Total	88	74	14	5.3:1
Race				
Chinese	72 (81.8%)	60	12	5:1
Malay	10 (11.4%)	9	1	9:1
Indian	4 (4.5%)	3	1	3:1
Others	2 (2.27%)	2	0	all female

Demographic data of Singapore

a) Population profile 1996[1]

Total population: 3,044,300

Racial distribution:

Chinese 77.3%
Malays 14.1%
Indians 7.3%
Others 1.3%

Sex ratio: 1013 males per 1000 females

Age below 15 years of age: 22.8% of population

b) 1996 age distribution[2]

Age (y)	n	(as a % of 0 to 14 years age group)
0 to 4	244,000	(35.2%)
5 to 9	241,500	(34.8%)
10 to 14	209,000	(30%)
15 to 19	203,000	

the age of 10 years (77.3%) with only 2 (2.3%) below the age of 5, and 18 (20.5%) between the ages of 5 and 10 years (Table II). The youngest was 4.5 years at diagnosis, the oldest almost 16, with the mean age of onset being 12 years.

Of the 88 patients, females numbered 74 (overall female to male ratio of 5.28:1). Below the age of 10 years, female predominance was less marked (3.2:1); above the age of 10 years the sex ratio approached adult ratios with a greater female preponderance of 6.44:1. The racial distribution and differing sex ratios in the different racial groups are shown in Table II.

The commonest presenting features locally were malar rash (54%), fever (40%), arthritis (35%), positive antinuclear antibody or ANA (88%) and double-stranded deoxyribonucleic acid or dsDNA (84%) (Table III). 13.6% had autoimmune haemolytic anaemia and 15.9% nephritis at presentation. To date, 26.1% have had neurological manifestations including fits, psychotic behaviour and focal deficits, 15.9% have had haemolytic anaemia, while 47.7% have had nephritis ranging from haematuria to nephritic or nephrotic syndrome and acute renal failure. Nine biopsy results available showed renal involvement of Class II (1 case), Class III (2 cases), Class IV (3 cases) and Class V (3 cases). For comparison, the initial manifestations in adult Asians with SLE are shown in Table IV.

Infection was a common complication including skin sepsis, abscess, tuberculosis, pneumonia, salmonellosis, shingles and septicaemia. Hypertension has involved 27% of the patients to date (compared to 28% cumulatively in western series). Raynaud's phenomenon (4.5%), thrombosis (4.5%), pulmonary embolism and infarct, pulmonary haemorrhage, angina and acute myocardial infarct and avascular necrosis of bone (only one patient each for all the rest) were rare in our children locally but they do occur. These patients were subsequently found to be positive for anti-cardiolipin antibody or lupus anticoagulant. Many also had other features of vascular occlusion such as fits or stroke.

Three children had a positive family history of SLE.

All have been on steroids and 44.3% needed cyclophosphamide or azathioprine, mainly for renal involvement. Twenty-seven per cent needed anti-hypertensives, 9% anticonvulsants and 9% lipid lowering agents.

Juvenile Chronic Arthritis (JCA)

JCA was the second commonest diagnosis for children with rheumatological disease under follow up after SLE, with pauciarticular JCA being the commonest and systemic JCA the rarest. The relative percentages of the different types of JCA locally is as follows: systemic onset (12.3%), pauciarticular onset (excluding SPA) (26.5%), polyarticular onset (20.4%) and spondyloarthropathy / AS (40.8%).

Still's disease (Systemic onset JCA): We have only a few patients with Still's disease (6); 4 were females and 2 were males. Age of onset ranged from 1.42 years to 13.42 years, averaging 6 years. Four have been complicated with prolonged flares requiring systemic steroids and methotrexate. All were independent in activities of daily living; none required joint replacement nor had any features of amyloidosis or haemophagocytic syndrome.

Pauciarticular onset JCA: Pauciarticular JCA (including SPA) was the commonest form of JCA with a male predominance (1.8:1). Excluding SPA, the male predominance remained at 1.6:1, the age at diagnosis ranged from 1.83 years to 16 years with a mean of 7.16 years. They comprised 11 Chinese (84.6%), 1 Malay (1.4%) and 1 Caucasian (1.14%). Nine had knee involvement, 5 had ankle involvement and 1 had hip involvement. Five (38.5%) were ANA positive, but none had evidence of uveitis on routine repeated slit lamp examination. All

TABLE III: MANIFESTATIONS OF SLE IN ASIAN CHILDREN AND CAUCASIAN CHILDREN[6]

	% at presentation		% cumulative	
	Asian, local	Caucasian[6]	Asian, local	Caucasian[6]
dsDNA	88			
ANA	84			
Malar rash	54	51	63	56
Fever	40			
Arthritis	35	72	50	76
Alopecia	30	16		
Cytopenia	28		68	
AIHA	14		18	
Thrombocytopenia	13	22	26	27
Oral ulcer	16	12	28	16
Nephritis	16	84	48	86
Photosensitivity	13	16	45	16
Neurological	9	9	26	31
Discoid rash	5		11	
Serositis	5	40	8	47

SLE: systemic lupus erythematosus; AIHA: auto-immune haemolytic anaemia; ANA: anti-nuclear antibody; DsDNA: double-stranded deoxyribonucleic acid

TABLE IV: INITIAL MANIFESTATIONS OF SLE IN CHINESE ADULTS IN SINGAPORE[3]

	%
Anti-DNA antibodies	100
Antinuclear antibodies	98
Skin and mucous membrane	52
Fever, malaise	48
Arthritis or arthralgia	44
Thrombocytopenic purpura	4
Neuropsychiatric	4
Haemolytic anaemia	3

SLE: systemic lupus erythematosus

had been on NSAIDs, 2 had intra-synovial steroid injections and 1 has undergone synovectomy for persistent localised synovitis.

Polyarticular onset JCA: We had 10 children with polyarticular JCA including one likely to have lupus overlap. This was a 10-year-old girl with rheumatoid factor (RF) positive symmetrical erosive arthritis as well as positive lupus serology but no other clinical features of lupus. At the time of study she was on methotrexate. There was a female preponderance of 1.5:1, an age range of 5.5 to 13.88 years at diagnosis and mean age of 9.8 years. Again the majority were Chinese. There were 8 (80%) Chinese with 1 Malay (10%) and 1 Indian (10%). One-third was RF positive (2 girls and 1 boy). All had been on NSAIDs, 4 (40%) had been on systemic steroids for a period, and one each were on methotrexate and sulphasalazine.

Spondyloarthropathy: There were a total of 20 patients with spondyloarthropathy, including 2 with radiographic changes of ankylosing spondylitis before the age of 16 (both Chinese boys aged 12.3 and 12.5 years at diagnosis), 1 with psoriatic arthritis and 1 with iritis. There was a male predominance with a male to female ratio of 2.3:1. They tended to be older at diagnosis than the other groups with an age range of 4.3 to 15.8 years and a mean age at onset of 10.5 years. Half of the patients needed second line drug therapy such as methotrexate or sulphasalazine. Three needed systemic steroids, 2 had intra-synovial steroid injections and 1 had undergone synovectomy. All were independent in activities of daily living and attending school or working.

Juvenile Dermatomyositis (JDM)

A total of 17 cases of juvenile dermatomyositis with a female predominance (4.6:1) were seen. The youngest was 2.8 years old at diagnosis. The average age of onset in males was younger (5.5 years) compared to females (7.7 years) with an overall mean age of onset of 7 years. The majority were Chinese (70.6%), with 17.6% Malays, 5.8% Indians and 5.8% of other races. All had skin involvement, ¾ had weakness, and ¼ developed calcinosis. There was only 1 case of dermatomyositis sine myositis. This was a 3-year-old girl with disease onset of 4 months. The majority needed steroids at some point during the course of their illness, one-third has been on hydroxychloroquine mainly for skin involvement.

Overlap Syndrome/MCTD

A total of 4 patients had features of overlap syndrome: 2 with features predominantly of lupus and scleroderma, 2 with features predominantly of lupus and dermatomyositis. All were females with an age range of 4 to 11 years at diagnosis. Three were Chinese and one Indian. None had renal manifestations. Disease in one was severe with vasculitis and recurrent infections, and milder in the remaining.[3]

Other Inflammatory Rheumatological Conditions (Polyarteritis nodosa, Scleroderma, Polychondritis, Chronic Recurrent Multifocal Osteomyelitis, Rheumatic fever)

One Malay boy with polyarteritis nodosa (PAN) presented at 14 years of age with fever, digital gangrene, oral ulcers and arthritis and responded to pulsed methylprednisolone. Renal and mysenteric angiogram were normal. Markers for lupus were negative.

A 6-year-old Chinese girl with polychondritis presented with recurrent stridor requiring tracheostomy. She also had swollen red ears and scleritis, and developed a saddle nose and arthritis involving the metacarpophalangeal joints, proximal interphalangeal joints, ankles, knees and xiphisternum. Disease was controlled with steroids, non-steroidal anti-inflammatory drugs (NSAIDs) and methotrexate.

One girl with systemic sclerosis presented at the age of 12.9 years with diffuse tightening of the skin, but no significant gastrointestinal, respiratory or renal complications. She was treated with steroids, NSAIDs and cyclosporin A.

A Chinese boy with Behcet's disease presented at the age of 14.5 years with oral and penile ulcers. He was treated with steroids and cyclosporin A.

Another Chinese boy presented with a lytic lesion at the proximal end of the left tibia. Biopsy showed nonspecific granulation which was culture negative. He subsequently developed involvement of the ankle and toe. Initial treatment empirically for infection was unsuccessful. He subsequently improved when treated with NSAIDs and physiotherapy.

One single case of rheumatic fever on follow up was a Malay boy with cardiac and joint involvement.

Five children were recently referred to our department for limb pains with hypermobility as the only definable abnormality. Their ages ranged from 2.4 to 7.9 years at diagnosis. None had Marfan's syndrome or Ehlers Danlos syndrome.

One Chinese boy was referred for a painful arm with abnormal posturing and great reluctance to move the extremity. The arm was cool, mottled, slightly swollen and severely painful with pain intensified by light touch. The onset was related to the onset of a family crisis. He was managed with family counselling and physiotherapy.

Discussion

The commonest disease for children under follow up was SLE, followed by JCA then JDM. Very few patients locally had the diagnoses of scleroderma, PAN, Behcet's disease or polychondritis probably because of the small size of Singapore's paediatric population.

The pattern locally of female predominance for lupus, JDM, overlap syndrome and polyarticular JCA, the pattern of male predominance for SPA, and the lack of gender difference for Still's disease are similar to Western data. A striking difference however lies in the male predominance and lack of uveitis in local children with pauciarticular onset JCA which is classically described as more common in girls and associated with uveitis. The racial distribution overall was similar to the general racial distribution.

SLE formed the largest group of patients in the paediatric registry still under follow up, probably because the disease is prolonged and has more complications. Those older than 16 years are under the care of adult rheumatologists, highlighting the importance of a continuity of care from childhood to adulthood.

The overall sex ratio of 5.28 females to 1 male locally is similar to the female predominance noted in the West of 4.5:1, although the overall ratio varies to some extent with age.[3-9] Female preponderance became more marked with increasing age from 3.2:1 to 6.4:1 to 9:1 (for ages less than 10 years, more than 10 years and adults respectively), a trend similar to Western data,[4-10] although one published report found no difference in sex ratio in lupus of onset before and after the age of 10 years.[11] The female preponderance was present in all the races but most marked in the Malays (9:1).

With regard to clinical features, malar rash and arthritis were similarly common presentations compared to Western data.[6] However, in Western children, arthritis and nephritis may be more common while photosensitivity and oral ulcers are more common locally. It is possible that with the strong sunshine in Singapore, photosensitivity is more noticeable here. Compared to adult onset SLE in local Chinese[3] (Table IV), similar figures are seen for arthritis or arthralgia, skin and mucous membrane involvement and fever. However, a greater percentage of children than adults have thrombocytopenia and haemolytic anaemia at presentation, and childhood lupus can present to the haematologist with pallor or bruising. A few children with disease complicated by angina and myocardial infarction, pulmonary hypertension, pulmonary embolism, Raynaud's phenomenon or fits, tested positive for anti-cardiolipin antibody or lupus anti-coagulant.

Patients with JCA comprise a relatively small proportion of the children with paediatric rheumatological conditions compared to SLE. This is possibly because JCA may be rarer locally or because the duration of follow up is shorter as the disease tends to be milder and less protracted. The majority of patients with JCA currently on follow up have been diagnosed recently (several months to a few years ago) unlike the patients with SLE some of whom have been on follow up for decades (12 have been on follow up for at least 10 years, while 5 have been on follow up for over 20 years). Pauciarticular JCA is the commonest and systemic onset JCA is the rarest, a pattern similar to many reported series.[12,13]

The small number of children with Still's disease is likely due to its rarity and our small population. The average age of onset is 6 years while published data report 1 to 4 years as the most common age of onset.[14-16] Although a few had disease which was difficult to control, none had been severely crippled by the disease, possibly because the disease is milder in Asians and our numbers are small.

The marked differences in pauciarticular JCA were the male predominance and lack of iritis. Locally, males predominate 1.6:1 unlike published data of female predominance of up to 7:1.[13] It is possible that some of our young males with pauciarticular onset disease may evolve into the syndrome of seronegativity, enthesopathy and arthropathy (SEA) later in life. The latter disease is generally more common in males and those with HLA B27 positivity. Of the 13 cases, 5 were ANA positive (38.5%) compared to 50% in Western series[13] but none of the cases have been found to have uveitis despite regular ophthalmologic assessment (compared to 40%[13]). This absence of eye involvement could be due to a difference in manifestations of the disease locally. On the other hand, it could be only an apparent difference because the population pool is smaller, the duration of regular assessment has been short, or cases diagnosed in previous years may not have had such regular eye assessments. Prolonged follow up will reveal if a significant proportion of males do indeed evolve into SEA, if any develop eye complications, or if our local population is indeed different. Genetic analysis will provide information on whether the genetic associations locally are different from Caucasian children. The commonest joints involved were the knees and ankles which is similar to published data.[17]

Children with polyarticular onset JCA had an average older age of onset than those with Still's disease or pauciarticular disease as well as a female predominance similar to Western studies.

There were more boys with spondyloarthropathy and ankylosing spondylitis locally, which is similar to Western studies. The disease was similarly more difficult to control than that in younger children with pauciarticular disease involvement, with half needing second line drug treatment.

For juvenile dermatomyositis, there was a similar female predominance,[18] average age of onset of 7 years, and younger average age of onset in males[19,20] as in Western reports. Clinical features were similar to those seen elsewhere.

Overlap syndrome was rare with only 4 cases.

Other inflammatory rheumatological conditions were rare with only one case each of PAN, polychondritis, scleroderma, Behcet's disease, chronic recurrent multifocal osteomyelitis and rheumatic fever. Rheumatic fever is rather rare now with improved socioeconomic status and health care in Singapore. Patients with mainly cardiac involvement are followed up by the cardiology service and were not entered into the rheumatology registry.

Hypermobility was the sole abnormality in 5 patients referred recently for limb pains. The small number is probably because some cases may have been managed by orthopaedic surgeons and also because few remain on follow up for long as the limb pain usually resolves with time.

The only child found to have reflex sympathetic dystrophy reflects the rarity of this condition locally.

Conclusion

Many childhood diseases may first present with a rheumatological complaint. This analysis of 170 patients currently on follow up for rheumatological disease gives an insight into the pattern of the disease locally, and shows the similarities and differences compared with Western data as well as local data on adults. SLE is the predominant disease with photosensitivity, malar rash, arthritis, infection, renal involvement and haemolytic anaemia being important features locally. Anti-phospholipid antibodies contributed to thrombotic events and significant morbidity. Children with JCA and JDM form a smaller proportion of the rheumatology pool. In our children with pauciarticular JCA, the male predominance and lack of uveitis are striking. Other conditions were rare.

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