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REVIEW ARTICLE

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Nomenclature and classification of juvenile idiopathic arthritis: where to after Durban?

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Abstract The Durban classification system for juvenile idiopathic arthritis (JIA) is reviewed in a historical context and in association with a review of problems that have become apparent. These include: (i) a family history of psoriasis as an exclusion factor (oligoarthritis and enthesitis-related arthritis) and as in inclusive factor (psoriatic arthritis); (ii) a family history of HLA-B27-associated disease as an exclusion factor (oligoarthritis) and an inclusive factor (enthesitis-related arthritis); (iii) the requirement of a dermatological opinion for psoriasis; (iv) the absence of HLA-B27 antigen in proband, with the presence of antigen in the family history; (v) the definition of time of onset; (vi) the presence of rheumatoid factor (RF) with oligoarthritis; (vii) HLA-B27-positive males with an onset of arthritis after 8 years of age. Modifications are suggested to maintain the homogeneity of groupings for research, whilst providing a practical scheme for clinicians. Three main modifications are suggested. (A) That the family history be included in descriptors rather than as inclusive or exclusion criteria. (B) Further development of the hierarchical system, which is partly used in the Durban classification. (C) That the following changes be made: rheumatoid-factor-positive oligoarthritis and polyarthritis be classified together; extended oligoarthritis and polyarthritis be classified together; HLA-B27-positive disease be classified with fewer inclusive and exclusion criteria; the criteria for psoriatic arthritis be modified; the classification of the disease in a particular child be changed in the event of relevant changes in the child's disease or laboratory profile. These suggestions are made to stimulate discussion.

Key words Juvenile idiopathic arthritis · Durban classification · Nomenclature · Juvenile rheumatoid arthritis

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Introduction

Paediatric rheumatology has a comparatively brief history, but from early times the problems of nomenclature and classification of arthritis in children were evident. In 1864, Cornil[1] wrote of a 29-year-old woman whose chronic arthritis had commenced at the age of 12. In 1890, Diamantberger[2] described a series of 38 cases of arthritis in children, most of whom were under the care of other physicians. It is the publication of George Frederic Still,[3] "On a form of chronic joint disease in children," in 1896 that is generally considered to mark the birth of paediatric rheumatology. The article included the first discussion of differing types of arthritis in children, and the need for a system of classification. Of his series of 22 children, he had personally cared for 19, and noted that there were at least three different types of arthritis. Twelve children were subject to recurrent pyrexia and had enlarged lymph glands and spleens with swollen joints, for which the term Still's disease was used. In the current terminology, these children would be classified as having systemic arthritis.

Approximately 50 years after this publication, the Special Unit for Juvenile Rheumatism[4] was established at Taplow, near London. The terminology used for childhood arthritis at this time varied from country to country.[5–8] In a study in 1962, Barbara Ansell defined Still's disease as "rheumatoid arthritis starting before the 16th birthday with evidence of arthritis in four or more joints for a minimum of three months or (in a few cases in which fewer than four joints were affected) with a biopsy of synovial membrane showing hyperplasia with underlying round-cell infiltration and plasma cells without evidence of tuberculosis or other infection".[9]

In Europe in 1977, the term juvenile chronic arthritis (JCA) as an umbrella term was first defined at a meeting in Oslo of the European League Against Rheumatism (EULAR) and was published the following year.[10] The EULAR criteria included the spondyloarthropathies. JCA was defined as arthritis present for more than 12 weeks with other conditions excluded. The term juvenile rheumatoid

arthritis (JRA) was used for the form of childhood arthritis where the rheumatoid factor was present in the blood of children, with multiple joints involved. In this system, the disease juvenile rheumatoid arthritis was considered separately from the term juvenile chronic arthritis, although in practice it was frequently considered to be one of the forms of JCA. Use of the term Still's disease gradually decreased over several decades, as the meaning of the term became less precise.

In the United States of America, there were parallel developments in the field of paediatric rheumatology. The American Rheumatism Association (ARA) was formed in 1932. In 1963, a committee was appointed by the ARA to study the possibility of a classification system for juvenile rheumatoid arthritis, and this was one of the first attempts at formal classification.

The ARA terminology was characterized by the umbrella term of "juvenile rheumatoid arthritis" (JRA),[10] which was defined as arthritis of at least 6 weeks duration with onset before the 16th birthday. The term JRA excluded the spondyloarthropathies (i.e., psoriatic arthritis and juvenile ankylosing spondylitis), but included cases where the rheumatoid factor was positive.

During the 1980s and early 1990s, the EULAR and ARA systems were both used extensively, but there were clear divisions between them (Table 1). In 1995, an international committee under the auspices of the World Health Organisation and the International League of Associations for Rheumatology (ILAR) met in Chile. Subsequently, a set of proposed criteria were published in 1995.[11] The stated aim of the proposal was to provide the framework for an internationally acceptable and applicable set of classification criteria for arthritis of childhood to be evaluated internationally and amongst a variety of ethnic populations. It was hoped that these criteria would facilitate more meaningful research and better patient care.

The proposed overall term was "idiopathic arthritis of children" (IAC), to replace the terms JCA and JRA. The definition was arthritis present for 6 weeks with onset before the 16th birthday with other diagnoses excluded. The seven subgroups included in this term were oligoarthritis, extended oligoarthritis, polyarthritis rheumatoid factor positive, polyarthritis rheumatoid factor negative, systemic arthritis, enthesitis-related arthritis, and psoriatic arthritis. This system maintained the arbitrary division of four or fewer affected joints for oligoarthritis, and more than four for polyarthritis. The term "pauciarticular" was eliminated.

A second meeting of the Classification Taskforce of the Pediatric Standing Committee of the International League of Associations for Rheumatology was held in Durban, South Africa, in 1997, and the proposed classification was published in 1998.[12] It was agreed at this meeting that the primary purpose of the classification was to facilitate communication among physicians and scientists. It acknowledged that it would be difficult for any such classification to fulfil multiple roles, including research, and communication with patients and community agencies.

The Durban classification defined juvenile idiopathic arthritis (JIA) as arthritis which was present for 6 weeks, with other conditions excluded, commencing before the 16th birthday and currently having no known cause. It defined six subgroups, one of which, oligoarthritis, was further subdivided into two groups after 6 months of disease, i.e., persistent or extended oligoarthritis, depending on the cumulative total of joints involved after the initial 6-month period. There was an additional category with two parts to accommodate disease that fitted the overall definition of JIA but did not fulfil the definition of any of the defined categories. This could also be used if the disease fitted two or more of the defined categories.

Each category in the proposed classification had defining criteria and listed descriptors, and five of the seven groups also had listed exclusions. It is known as the Durban classification, and is given in detail below.

Table 1. Summary of the differences between juvenile chronic arthritis (JCA) and juvenile rheumatoid arthritis (JRA) and the defined subgroups of each

	EULAR criteria: JCA, arthritis commencing before 16th birthday with other conditions excluded	ARA criteria: JRA, arthritis commencing before 16th birthday with other conditions excluded
Duration of arthritis necessary for diagnosis	12 weeks	6 weeks
Polyarticular onset, rheumatoid factor (RF)-positive arthritis: onset of disease in more than 4 joints with RF present	Alternative 1: subgroup of JCA. Alternative 2: in some studies using EULAR criteria, this group of patients is diagnosed with JRA, and is considered to be outside the definition of JCA	Subgroup of JRA
Juvenile ankylosing spondylitis: arthritis initially in a few joints associated with the presence of HLA-B27, and most common in older boys and in lower limb joints	Subgroup of JCA	Not included in the term JRA; considered to be a separate condition
Psoriatic arthritis: arthritis and psoriasis	Subgroup of JCA	Not included in the term JRA; considered to be a separate condition
Arthritis associated with inflammatory bowel disease	Subgroup of JCA	Not included in the term JRA; considered to be a separate condition

Durban classification

Systemic arthritis

Definition

Arthritis with or preceded by fever of at least 2 weeks duration that is documented to be quotidian for at least 3 days, and accompanied by one or more of the following symptoms:

1. evanescent, nonfixed, erythematous rash,
2. generalized lymph node enlargement,
3. hepatomegaly or splenomegaly,
4. serositis.

Descriptors

1. Age at onset.
2. Pattern of arthritis during the onset period (i.e., during the first 6 months of disease): (a) oligoarthritis, (b) polyarthritis, (c) arthritis present only after 6 months of systemic illness.
3. Pattern of arthritis during the disease course (i.e., after the first 6 months of disease): (a) oligoarthritris, (b) polyarthritis, (c) no arthritis after the first 6 months of systemic disease.
4. Features of systemic disease after 6 months.
5. Presence of rheumatoid factor (RF).
6. Level of C-reactive protein.

Oligoarthritis

Definition

Arthritis affecting 1–4 joints during the first 6 months of disease. Two subcategories are recognised:

1. *Persistent oligoarthritis*; which affects no more than 4 joints throughout the disease course.
2. *Extended oligoarthritis*; which affects a cumulative total of 5 joints or more after the first 6 months of the disease.

Exclusions

1. Family history of psoriasis confirmed by a dermatologist in at least one first or second degree relative.
2. Family history consistent with medically confirmed HLA-B27 associated disease in at least one first or second degree relative.
3. Positive RF test.
4. HLA-B27 positive male with onset of arthritis after 8 years of age.
5. Presence of systemic arthritis as defined above.

Descriptors

1. Age at onset of arthritis and psoriasis.
2. Pattern of arthritis at 6 months and last clinic visit:
   (a) large joints only,
   (b) small joints only,
   (c) limb predominance: (i) upper limb predominant, (ii) lower limb predominant, (iii) no upper or lower limb predominance,
   (d) specific joint involvement (e.g., hip, cervical spine, etc.),
   (e) symmetry of arthritis.
3. Occurrence of anterior uveitis (acute or chronic).
4. Presence of antinuclear antibody (ANA).
5. HLA class I or II predisposing or protective alleles.

Polyarthritis (rheumatoid factor negative)

Definition

Arthritis affecting 5 or more joints during the first 6 months of disease; tests for RF negative.

Descriptors

1. Age at onset of arthritis.
2. Symmetry of arthritis.
3. Presence of ANA.
4. Occurrence of uveitis (acute or chronic).

Polyarthritis (rheumatoid factor positive)

Definition

Arthritis affecting 5 or more joints during the first 6 months of disease, associated with positive rheumatoid factor tests on 2 occasions at least 3 months apart.

Exclusions

1. Absence of positive tests for RF on 2 occasions at least 3 months apart.
2. Presence of systemic arthritis defined as above.

Descriptors

1. Age at onset.
2. Symmetry of arthritis.
3. Presence of ANA.
4. Immunogenetic characteristics comparable to adult populations with rheumatoid arthritis.

Psoriatic arthritis

Definition

1. Arthritis and psoriasis, or
2. arthritis and at least 2 of:
   (a) dactylitis,
   (b) nail abnormalities (pitting or onycholysis),
   (c) family history of psoriasis confirmed by a dermatologist in at least one first-degree relative.

Exclusions

1. Presence of RF.
2. Presence of systemic arthritis as defined above.

Descriptors

1. Age at onset of arthritis or psoriasis.
2. Pattern of arthritis 6 months after disease onset, and at last clinic visit:
   (a) large joints only,
   (b) small joints only,
   (c) limb predominance: (i) upper limb predominant, (ii) lower limb predominant, (iii) no upper or lower limb predominance,
   (d) spinal involvement,
   (e) sacroiliac joint involvement,
   (f) glenohumeral joint involvement,
   (g) hip joint involvement,
   (h) sternoclavicular joint involvement,
   (i) symmetry of arthritis.
3. Disease course:
   (a) oligoarthritis,
   (b) polyarthritis.
4. Presence of ANA.
5. Anterior uveitis (specify):
   (a) chronic anterior uveitis,
   (b) uveitis that is characterized by pain, redness, or photophobia.
6. HLA descriptors.

Enthesitis-related arthritis

Definition

1. Arthritis and enthesitis, or
2. arthritis or enthesitis with at least 2 of:
   (a) sacroiliac joint tenderness and/or inflammatory spinal pain,
   (b) presence of HLA-B27,
   (c) family history in at least one first- or second-degree relative of medically confirmed HLA-B27-associated disease,
   (d) anterior uveitis that is usually associated with pain, redness, or photophobia,
   (e) onset of arthritis in a boy after the age of 8.

Exclusions

1. Psoriasis confirmed by a dermatologist in at least one first- or second-degree relative.
2. Presence of systemic arthritis as defined above.

Descriptors

1. Age at onset of arthritis or enthesitis.
2. Pattern of arthritis at 6 months and at last clinic visit:
   (a) large joints only,
   (b) small joints only,
   (c) limb predominance: (i) upper limb predominant, (ii) lower limb predominant, (iii) no upper or lower limb predominance,
   (d) spinal involvement,
   (e) sacroiliac joint involvement,
   (f) glenohumeral joint involvement,
   (g) hip joint involvement.
3. Symmetry of arthritis.
4. Disease course:
   (a) oligoarthritis,
   (b) polyarthritis.
5. Presence of inflammatory bowel disease.

Other arthritis

Definition

Arthritis in children of unknown cause persisting for at least 6 weeks that:

1. does not fulfil criteria for any of the other categories,
2. fulfils criteria for more than one of the other categories.

Terms used in this system of classification

Glossary terms

Arthritis. Swelling within a joint, or a limitation in the range of joint movement with joint pain or tenderness, which persists for at least 6 weeks, is observed by a physician, and which is not due to primarily mechanical disorders (also see exclusion list in ACR criteria for juvenile arthritis).
Number of affected joints. Joints able to be individually evaluated clinically to be counted as separate joints.
Sacroiliac joint arthritis. Presence of tenderness on direct compression over the sacroiliac joints.
Quotidian fever. Daily recurrent fever that rises to 39°C or above once a day and returns to 37°C or below between fever peaks.
Serositis. Pericarditis, pleuritis, and/or peritonitis.
Enthesitis. Tenderness at the insertion of a tendon, ligament, joint capsule, or fascia to bone.
Psoriasis, family history of psoriasis. Diagnosed by a dermatologist.
HLA-B27-associated disease. Ankylosing spondylitis; sacroiliitis with inflammatory bowel disease; acute (symptomatic) anterior uveitis.
Dactylitis. Swelling of one or more digits, usually in an asymmetric distribution, that extends beyond the joint margin.
Positive test for rheumatoid factor. At least 2 positive results (as routinely defined in a laboratory using the WHO standard), 3 months apart, during the first 6 months of observation.

Nail pitting. A minimum total of 2 pits on one or more nails at any time.
Inflammatory spinal pain. Pain in the spine at rest, with morning stiffness in the spine that improves with movement.
Uveitis. As diagnosed by an ophthalmologist.
Spondyloarthropathy. Inflammation of entheses and joints of the lumbosacral spine.

Descriptor terms

Features of systemic disease. Fever, rash, serositis, hepatomegaly, splenomegaly.
HLA Class I or II predisposing or protective alleles. Description of alleles positively or negatively associated with the category.
Limb predominance. Description of any definite predominance of arthritis in upper or lower limbs
Positive test for antinuclear antibody. At least 2 positive results (as routinely defined in the laboratory you use) 3 months apart, during the first 6 months of observation with technique and titer indicated.
Oligoarthritis. Arthritis in a cumulative total of 1–4 joints.
Polyarthritis. Arthritis in a cumulative total of more than 4 joints.
Large joints. Hip, knee, ankle, wrist, elbow, glenohumeral joints.
Small joints. All others.
Symmetry. A predominantly symmetrical or predominantly asymmetrical pattern of joint involvement.

In summary, the proposed Durban classification provided greater clarification of subgroups with six main categories, one having two parts, and a seventh additional category for disease that did not fit exclusively into one of the other categories. There was a descriptive framework of the disease, in addition to defining features for each disease category. There were practical considerations such as the need to commence treatment reasonably early in systemic arthritis rather than leaving fever untreated for 2 weeks for the purpose of documentation.

This proposed Durban classification system, as with all systems of classification, brings into focus the dichotomy between the needs of researchers and the needs of clinicians. The work of each is essential for the other, and a classification system should be acceptable to both.

Problems of the Durban classification

In evaluations of the Durban criteria, several significant problems are becoming apparent. Some of these are listed below.

Family history of psoriasis as an exclusion factor

The issue of family history has consistently created difficulties. For two subgroups, oligoarthritis and enthesitis-related arthritis (ERA), listed exclusions include a family history of psoriasis in first- or second-degree relatives. It follows that if it is not possible to define the presence or absence of psoriasis diagnosed by a dermatologist in all first- or second-degree relatives, the diagnosis of oligoarthritis or enthesitis-related arthritis should not be made. The disease would then be categorized as "other" arthritis. This would include the disease of any adopted child, any child of unknown or doubtful paternity, or any child whose extended family is no longer in contact with the patient's immediate family. It is known that the paternity of children in normal families may well not be as expected, and the percentage of children fathered outside of a marriage is significant and frequently not defined. Thus, the requirement to know the family history of psoriasis causes a significant numbers of cases of juvenile idiopathic arthritis to become classified as "other" arthritis. In addition, it causes inaccuracies of unknown extent because of paternity issues.

An example of this problem is the case of a girl of 4 years of age with JIA who presented with both ankles swollen. She was adopted. The family history was not known, and in particular there was no history regarding psoriasis. She had no evidence of psoriasis. She was ANA-positive, RF-negative, and HLA-B27-negative. Her disease was categorised as "other" arthritis, although it is likely that she had oligoarthritis.

Family history of psoriasis as an inclusive criteria

A family history of psoriasis is included as one of three possible criteria, of which two are required, along with arthritis, for the diagnosis of psoriatic arthritis. Thus, children for whom the family history is unknown or incomplete may have a disease that would fulfil the criteria for psoriatic arthritis if their complete history was known.

An example of this problem is the case of a boy of 5 years of age who presented with arthritis consistent with JIA, and with a swollen knee and ankle. He had six pits in his fingernails, but no evidence of a psoriatic rash and was otherwise clinically normal. He was adopted and there was no known family history. His JIA was classified as "other" arthritis, although it is most likely to be consistent with psoriatic arthritis.

In the Durban system, there is also an inconsistency where a family history of psoriasis is required in a first-degree relative as one possible inclusive criteria for psoriatic arthritis, and yet if psoriasis is present in the family history of a first- or second-degree relative, this is an exclusion factor for oligoarthritis. This may lead to more cases being classified as "other," where the disease is excluded from oligoarthritis, but not included in psoriatic arthritis.

An example of this problem is the case of a boy of 3 years of age who had arthritis consistent with JIA in one elbow and both knees. A second-degree relative had psoriasis confirmed by a dermatologist. The boy did not have psoriasis, dactylitis, nail changes, HLA-B27 antigen, enthesitis, or

spinal pain, etc. This boy's disease was excluded from both oligoarthritis and psoriatic arthritis, and would be classified as "other" arthritis.

Family history of HLA-B27-associated disease

A family history of HLA-B27-associated disease is an excluding factor for the diagnosis of oligoarthritis and is a possible inclusive factor for ERA. Thus, ankylosing spondylitis, symptomatic anterior uveitis, and sacroiliitis with inflammatory bowel disease in relatives must be sought in the family history. It is known that these conditions may be slow to be diagnosed, or may never be. Thus, to ensure that these conditions have been excluded in relatives, it may be necessary for the relatives to be checked for the presence of disease and for the HLA-B27 antigen. If this cannot be done, the child's disease should be classified as "other" arthritis. Two examples of this problem are given below.

(i) A girl of 2 years of age had 2 swollen knees for more than 6 weeks, consistent with JIA. She was RF-negative and HLA-B27-negative. She was from a large family spread around the world, for whom little family history was available, and in whom HLA-B27-associated disease could not be excluded. Her arthritis was categorised as "other," although it is most likely to be consistent with oligoarthritis.

(ii) A boy of 6 years of age presented with a swollen knee and a swollen ankle which had persisted for at least 6 weeks, consistent with JIA. He was positive for HLA-B27. He did not have back pain or evidence of enthesitis. His paternal uncle was known to have a stiff back, which he considered was work-related, but the problem had never been investigated, and testing for HLA-B27 was not possible because of the uncle's location. This child's disease could not be classified as oligoarthritis because the exclusion criteria have not been clarified; nor could it be classified as ERA because there are insufficient criteria. His disease was classified as "other" arthritis, although it is most likely to be consistent with ERA.

Family history and a dermatological opinion

The classification system is further complicated by the requirement for a dermatologist's opinion for a diagnosis of psoriasis in the patient or a relative. This is relevant to the subgroups of oligoarthritis and ERA with psoriasis listed in the exclusions, and for the subgroup of psoriatic arthritis, where psoriasis is one of the inclusive criteria. Whilst it would be ideal to have a dermatologist's opinion in every possible case, this is unrealistic, particularly in the case of all second-degree relatives. It may not be known whether a dermatologist has been involved, particularly where the relative has died or is no longer in contact with the patient's immediate family. Thus, disease in a significant number of children may not be classifiable because of the requirement of a dermatologist's input. The category of "other" would again be the correct subgroup for the disease in such children.

An example of this problem is the case of a girl of 5 years old who presented with 2 swollen knees which had persisted for at least 6 weeks, consistent with JIA. She was HLA-B27-negative and RF-negative. The child had no evidence of psoriasis, but the maternal aunt was said to have had psoriasis as a child and young woman. It was not known whether a dermatologist had diagnosed the condition. The aunt's whereabouts were not known. Hence this child's disease was correctly classified as "other," because a dermatologist had not definitely diagnosed psoriasis in the aunt.

Family history positive, with an absence of HLA-B27 in the proband

The Durban criteria do not specify that if the proband is HLA-B27-negative, oligoarthritis can be diagnosed even though a relative has proven HLA-B27-associated disease. Thus, any first- or second-degree relative of any person who has HLA-B27-positive ankylosing spondylitis, acute anterior uveitis, or sacroiliitis with inflammatory bowel disease is excluded from being diagnosed with oligoarthritis.

An example of this problem is the case of a boy of 6 years of age who was HLA-B27-negative and RF-negative, and presented with JIA involving one knee which had persisted for 7 months. He had an uncle with HLA-B27-positive ankylosing spondylitis. This boy could not be classified as having oligoarthritis, although his disease would otherwise fit this category. His disease was classified as "other."

Definition of time of onset

For the Durban classification, it is necessary to define the time of onset of disease in order that the course of the disease in the first 6 months can be documented, i.e., how many joints are involved in the first 6 months. This is essential information, according to the criteria, for the differentiation between extended oligoarthritis and polyarthritis. However, it is often not possible to determine when the disease started or how many joints were involved at the onset. A series of 42 consecutive new cases of JIA, diagnosed in the setting of a modern health care system in Australia,[13] should that the mean duration from estimated time of onset to time to diagnosis was 40 weeks. Thus for some of these 42 children it would not have been possible to differentiate with certainty between polyarthritis and extended oligoarthritis. It may be argued that a diagnosis of polyarthritis using the Durban classification can only be made if there is definite evidence that the disease was not present 6 months prior to the first diagnosis.

An example of this problem is the case of a girl who was 7 years of age at the time of first diagnosis of JIA, had 2 swollen knees, 2 swollen ankles, and 2 swollen small joints in her fingers. One leg was significantly longer than the other, suggesting that arthritis had started earlier in one of the knees. The child was known to have had an abnormal gait for a number of years, suggesting that arthritis had been

present but undiagnosed for years. Her music teacher finally noticed the swollen finger joints and suggested that the parents should seek medical help. This child's disease is classified as "other" arthritis because of the missing information for the first 6 months of the disease, preventing differentiation between extended oligoarthritis and polyarthritis.

The current definition of RF-positive polyarthritis requires the tests to be done in the first 6 months of the disease. If the child is not seen in the first 6 months of the disease, or if it is not known when the disease started, the disease in some children would be classified as "other" when it is consistent with polyarthritis and RF has been shown to be positive, but not necessarily within the first 6 months.

The presence of RF with oligoarthritis

A further difficulty in the Durban system is when disease affects fewer than 5 joints, and rheumatoid factor is present. Currently, this disease would be classified as "other" arthritis.

HLA-B27-positive male with onset of arthritis after 8 years of age

Currently, arthritis in a male with onset after 8 years of age and with HLA-B27 present, but with no enthesitis, spinal pain, sacroiliac tenderness, relevant family history, or anterior uveitis, would be classified as "other" arthritis.

In summary, using the proposed Durban criteria, no child can be accurately diagnosed with oligoarthritis or ERA unless there is complete and accurate knowledge of the past and present health of first- and second-degree relatives in relation to psoriasis, acute anterior uveitis, sacroiliitis, inflammatory bowel disease, and ankylosing spondylitis. It requires that a dermatologist has been able to review all possible psoriatic rashes, and that all relatives have been tested for HLA-B27 where there are relevant symptoms of the above conditions. Likewise, no child with an incomplete family history, who has arthritis and either psoriatic nail changes or dactylitis but no psoriatic rash, can be diagnosed as having psoriatic arthritis. The diagnosis of "other" arthritis would be made where there remains any lack of information on any of these matters. In addition, if the exact time of onset is not well documented, no differentiation can be made between extended oligoarthritis and polyarthritis, and the classification "other" would be required.

Review of the literature

Some of the international evaluations of the Durban classification are now reported. In the Italian study by Fantini,[14] difficulties listed include the family history of psoriasis, the definition of numbers of joints involved in the first 6 months, and confirmation by a dermatologist of a diagnosis of psoriasis. A hierarchical system with the following order was suggested: systemic, rheumatoid factor positive polyarthritis or oligoarthritis, spondyloarthritis (ERA and psoriatic arthritis), oligoarthritis, and polyarthritis (RF-negative). In this system, a further category, rheumatoid factor positive oligoarthritis, would have to be created. Currently, this type of JIA is rare, but has no assigned category which would allow multicenter studies of this particular subgroup of JIA. The suggested hierarchical system is practical for clinicians, and echoes to some degree the Durban classification's system of exclusions.

In Cassidy's[15] evaluation of the Durban criteria undertaken at the University of Missouri, 54 children originally diagnosed with oligoarthritis by ACR criteria were re-evaluated to determine how many met the ILAR criteria for psoriatic arthritis or spondyloarthropathy. It was considered that there were no such children.

In a German study of the Durban classification, Foeldvari[16] showed that 12% of 97 patients were classified in the "other" arthritis category. The family history of psoriasis proved to be the most significant problem in making a more specific diagnosis.

In a British study by Thomson et al.[17] concerning the ILAR criteria for 502 Caucasian JIA patients, the alleles DRB1*11 were associated with JIA, and DRB1*08 and DPB1*0201 were associated with some subgroups.

In the Norwegian study by Flato et al.,[18] 13% of 337 patients were classified in the "other" category, with the main problem being the family history of psoriasis. The classification was studied in relation to outcome measures, and it showed the subgroups to have more homogenous outcomes than previous classification systems.

In a British study by Cleary et al.,[19] of 39 children with JIA and who were positive for HLA-B27, 10% could not be classified, mostly because of the family history of psoriasis.

An interesting idea for classification by latent class analysis has been proposed by Barrett et al.,[20] and this was applied to 540 children in a British study. It uses sophisticated mathematical procedures, and requires large numbers of children.

Discussion

For research purposes homogenous disease types are ideal, and everything possible should be done to ensure maximum homogeneity by excluding disease types that do not exactly fulfil the relevant criteria. Clinicians know that many children do not exactly fulfil specified criteria, but each of them deserves the best possible care. Overall, the Durban classification is more specific in its subgroups than previous classifications, and therefore research groups may be better defined, but clinicians have significant problems, with many children having a disease that fits only in the category of "other" where the proposed classification has been strictly followed.

Suggestions for modifications to the Durban system

Some suggested modifications to the Durban proposal have been put forward.

1. Family history may be pertinent in predicting the outcome of disease in some children, and it is certainly important in defining homogenous groups. In order to improve the classification, it is suggested that family history be taken out of the inclusive and exclusion criteria, and that it be recorded in the descriptors for each group. Thus for research purposes, descriptions of homogenous groups would be available and defined not only by criteria, but also by descriptors, including family history. This information would then be available for research purposes, but its absence it would not preclude the child's disease from being classified into one of the definite groups, and not as "other" arthritis. As the family history changes or becomes known, so would the descriptors for a particular child's disease. Thus, the homogeneity required for research would be constantly redefined and updated, and at the same time clinicians would have the benefit of a practical classification system.
2. The current Durban system is partly hierarchical. It is suggested that the hierarchical system should be developed further (details below).
3. It is suggested that the following measures should also be adopted: (i) RF-positive oligoarthritis be classified with polyarthritis, and be differentiated in the descriptors; (ii) extended oligoarthritis and polyarthritis be classified together, with the difference being defined in the descriptors as far as possible on the disease course; (iii) the presence of HLA-B27 be given greater importance except for systemic arthritis and possibly RF-positive arthritis; (iv) a disease is changed from one category to another when (and if) it becomes apparent that a feature of the disease or of the family history has changed. This would include disease which is originally in fewer than 5 joints and classified as oligoarthritis, but where more than 5 joints are involved after 6 months. The disease would then be classified as polyarthritis. The course of the disease would be included in the descriptors, and thus homogeneity for research purposes would be maintained.
4. It is suggested that the following minor changes in wording or definitions be made: (i) "evanescent, nonfixed" be changed to "evanescent"; (ii) "the first 6 months of the disease" be removed from the definition of polyarthritis (rheumatoid factor positive); (iii) "anterior uveitis that is usually associated with pain, redness, or photophobia" in the definition of ERA be changed to "uveitis associated with pain, redness, or photophobia" to maintain consistency with descriptors in psoriatic arthritis; (iv) deletion of "usually in an asymmetric distribution" from the definition of "dactylitis"; (v) the definition of "small joints" be changed to exclude spine and sacroiliac joints; (vi) deletion of "is observed by a physician" from the definition of "arthritis"; (vii) "in the laboratory you use" be changed to "in the particular laboratory" for the descriptor term for ANA; (viii) "Pericarditis, pleuritis, and/or peritonitis" in the definition of "serositis" be changed to "Pericarditis, pleuritis, or peritonitis."

Suggested hierarchical system

The suggested hierarchy is listed in order below. It should be noted that not all details have been listed; many would remain largely as in the Durban classification. Naming of these subgroups may need to be modified, which is beyond the scope of this article. Descriptors for each subgroup should include family history, course, number of joints, relevant laboratory parameters, associated features, and any information that is considered to be relevant to the defining of homogenous groups.

1. Systemic arthritis with no exclusions.
2. Rheumatoid factor positive JIA, irrespective of the number of joints involved, with the number of joints noted in the descriptors. The rheumatoid factor would need to be positive twice within a defined period, but not necessarily within the first 6 months of disease or of observation. The exclusion would be systemic arthritis.
3. Psoriatic arthritis, being arthritis and psoriasis, or arthritis and both dactylitis and nail changes consistent with psoriasis. The descriptors would include the number of joints affected, the presence of a family history of psoriasis or HLA-B27-associated disease, and the presence of HLA-B27 in the child and any known relatives. The exclusions would be systemic arthritis and the presence of RF.
4. ERA with descriptors to record the number of joints involved, the presence of enthesitis, sacroiliac tenderness, spinal pain, and family history (if possible) regarding HLA-B27-associated disease and psoriasis and whether a dermatologist made the diagnosis. Exclusions to this group would include systemic arthritis, the presence of RF, psoriasis or a combination of dactylitis and psoriatic nail changes. If psoriasis or a combination of dactylitis and psoriatic nail changes should later develop in the child, the disease would be reclassified as psoriatic arthritis. It is suggested that a new label be coined for this subgroup with "enthesitis" removed, because in many countries, enthesitis is moderately rare. It is noted that the use of "HLA-B27" would not be appropriate in the label. Although the presence of this antigen would be mandatory for defining the subgroup, it may also be present in systemic arthritis, RF-positive arthritis, and psoriatic arthritis, which are first excluded prior to classification into this subgroup.
5. Rheumatoid factor negative polyarthritis (more than 5 joints involved), with the number of joints involved in the first 6 months (if known) and the number involved later, to be recorded in the descriptors. This subgroup would include the current subgroup of extended oligoarthritis. Exclusions would be systemic arthritis, the

presence of RF, psoriasis or a combination of dactylitis and psoriatic nail changes, and the presence of HLA-B27.

6. Rheumatoid factor negative oligoarthritis (fewer than 5 joints involved). Exclusions would be systemic arthritis, the presence of RF, psoriasis or a combination of both dactylitis and psoriatic nail changes, and the presence of HLA-B27. In the descriptors, the presence of a family history of psoriasis would be recorded, and likewise the possible involvement of a dermatologist. If psoriasis, or the combination of dactylitis and nail changes, or involvement of more than 5 joints should develop later, the child's disease would be reclassified as psoriatic arthritis or polyarthritis, respectively (Fig. 1).

In summary, modifications to the Durban criteria are proposed whereby homogenous groups are defined by a mixture of inclusive and exclusion criteria and descriptors, including family history and disease course in the first 6 months and thereafter. It is suggested the hierarchical system is further developed, and that subgrouping is simplified to combine extended oligoarthritis with polyarthritis, and to combine oligoarthritis and polyarthritis were RF is positive. Thus, all appropriate detailed information would be available for research purposes. For clinical purposes, classification would be simplified and within a system that is largely hierarchical, but which has a degree of flexibility if disease features should change. The suggested system overcomes the problem that disease is unclassifiable without detailed information about the family history and of the first 6 months of the disease, including blood tests and joint count. It provides categories for a disease such as RF-positive oligoarthritis, which is rare but important.

Conclusion

In the suggested system, the dual purpose of a classification system would largely be addressed, i.e., maximum homogeneity for research purposes with a practical scheme for optimal clinical care.

It is only through debate and research that the classification and nomenclature of childhood arthritis can be improved. It is hoped that the suggestions presented herein will help to stimulate both debate and research, and ultimately help in the care of children with arthritis.

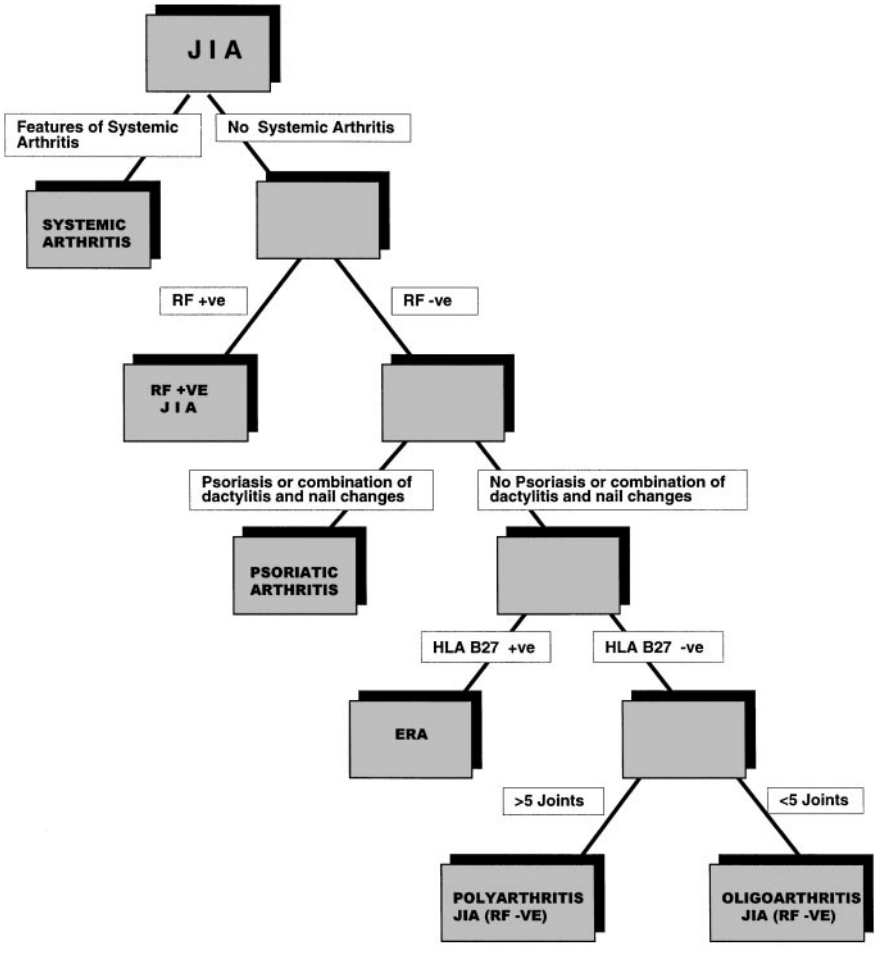

Fig. 1. Suggested hierarchal classification of juvenile idiopathic arthritis. JIA, juvenile idiopathic arthritis; RF, rheumatoid factor; +ve, positive; −ve, negative; ERA, enthesitis-related arthritis

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