Juvenile Rheumatoid Arthritis

By Dr. Wint Wint Tun. Edited by Ib Leth Nielsen, DDS, MSc

Orthodontic programs in the United States are often asked to take on post-graduate dental students in addition to their regular residents; these are often international students interested in spending time in a program of their choice in order to learn more about orthodontics and research.

At UCSF, we have listened to these requests and created two types of programs. One is a short-term program of three months' duration that we call a Preceptorship. The second is an International Fellowship Program in Orthodontics; it is of one year's duration, involves a more focused research endeavor, and is intended for young faculty in overseas dental schools who have a serious interest in an academic career, as well as experienced clinicians who have a couple of years in practice but want to expand their knowledge. In the past, we have been very successful with similar arrangements, and several fellows have later become Chairs at their respective universities overseas. In this issue of the Bulletin, *we have asked one of our recent Preceptors, Dr. Wint Wint Tun from Burma, to present her study of juvenile rheumatoid arthritis (JRA) as an example of the kind of limited research projects we expect our international students to undertake during their tenure.*

—I. L. Nielsen

INTRODUCTION

JRA is the most common chronic rheumatologic disease in children, and one of the most common chronic diseases of childhood. The overall prevalence of JRA is estimated to be from 30 to 150 per 100,000 children. In the United States and Canada there are an estimated 30,000 to 60,000 children and adolescents with the disease.[1]

JRA is characterized by chronic inflammation of the synovium and presence of articular cartilage damage.

In patients with JRA, the prevalence of clinically detectable temporomandibular joint (TMJ) involvement varies between 38% and 72%, depending on the diagnostic method used and the JRA type.[2]

According to the American College of Rheumatology (ACR) pediatric criteria for JRA, the disease is classified into three groups (Figure 1). An example of typical facial features of JRA is shown in Figures 2, 3, 4, 5, and 6.

JRA

Systemic JRA
- 20% of JRA patients
- Affects males and females equally
- Arthritis, high concomitant fever and rheumatoid rash
- Involves small joints of hands, wrists, knees and ankles
- May have internal organ involvement: hepatosplenomegaly

Polyarticular JRA (Poly)
- 40% of JRA patients
- More common in females
- Five or more joints affected in first six months of disease
- Involves large and small joints of legs and arms as well as jaw and neck
- Symmetrical distribution

Pauciarticular JRA (Pauci)
- 40% of JRA patients
- Common in females under eight years of age
- Four or fewer joints affected in first six months of disease
- Involves large joints: knees, ankles or wrists
- Asymmetrical distribution

Figure 1. The three different types of JRA.

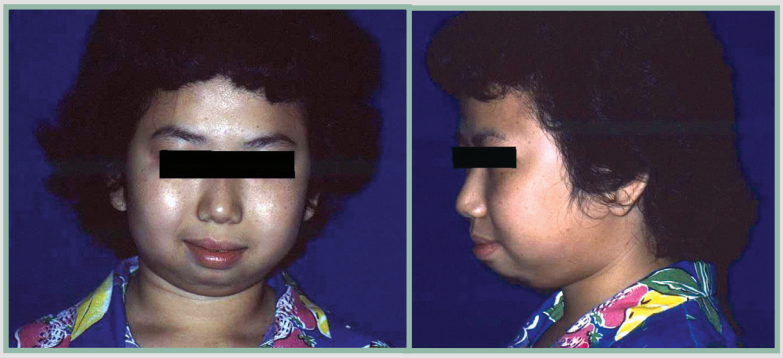

Figure 2. This 16-year-old patient diagnosed with the systemic type of JRA shows typical facial features of JRA.

Figure 3. Frontal view of teeth in occlusion showing anterior open bite and lower crowding.

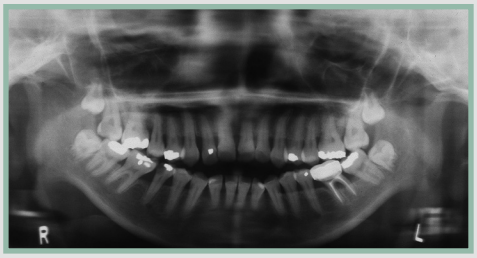

Figure 4. Panorex of the patient in Figures 2 and 3, showing condylar flattening in both right and left TMJ.

Figure 5. Lateral headfilm of the patient in Figure 2.

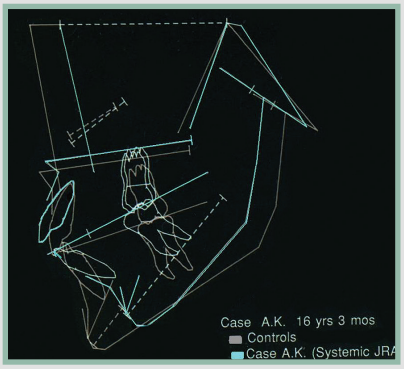

Figure 6. Facial morphology of the patient in Figure 2. Note the steep mandibular plane angle and convex profile. Patient and controls superimposed on nasal sella line.

Several studies have examined the facial morphology of JRA patients by means of lateral cephalograms. Some of the important findings include posterior inclination of the mandible in relation to the cranial base, resulting from posterior rotation due to condylar resorption during growth; also, a retrognathic mandible and reduction in overall mandibular dimensions are typical findings.[3,4,5] The changes in the mandible were clearly related to condylar damage due to temporomandibular joint arthritis.[6] Despite considerable agreement on the facial morphology in these patients, there is still limited literature on disturbances in skeletal growth, with most of it based on individual case reports.

OBJECTIVE

The objective of this study was to examine and compare the facial morphologies of the three different types of JRA patients cephalometrically. The second aim was to compare these findings to those in healthy children of the same age and sex. The third aim was to investigate the possible effects of JRA on skeletal development and maturation by means of the Tanner and Whitehouse TW2 skeletal age assessment method.[7]

SUBJECTS AND METHODS

A total of 15 JRA patients (Figure 7) were studied. These patients were at the time under treatment at Valley Children's Hospital in Fresno, CA.

	Sex		Age (in years)		
	Girls	Boys	Minimum	Mean	Maximum
Systemic	4	3	6.8	12	16.9
Poly	4	1	3.10	12	14.9
Pauci	1	2	7.6	12	15.9

Figure 7.

The facial morphology of JRA patients was analyzed using conventional lateral cephalometric headfilms. Cephalometric landmarks were identified and digitized on each patient's radiograph using the cephalometric analysis software Tiops™. The cranial, sagittal, vertical and dento-alveolar measurements of each patient were recorded, and the mean values and standard deviations were compared to those of normal children of the same age (control group) as available in the Tiops program database.

To evaluate the patients' skeletal development, the TW2 RUS method (Tanner, Whitehouse) was used. This method scores 13 regions of interest on each hand-wrist X-ray. The resulting scores were added to obtain the overall skeletal age, and the result was compared to the chronological age of normal children of the same age and sex. To ensure the accuracy of the stage of maturation determined visually, all hand-wrist films were also scanned and analyzed automatically by a new program BoneXpert™. [8] The results showed great agreement between the two methods.

RESULTS

In the group with systemic JRA, the S-N-B (Figure 8) and S-N-Pg (Figure 9) angles were significantly reduced, indicating a true mandibular retrognathia with an associated increase in the sagittal jaw relationship A-N-Pg (Figure 10).

The increase in mandibular plane angle (ML/MRLar) (Figure 11) indicates that mandibular plane is steeper than normal in this group. The reduced ML/MBLar angles, describing the shape of the mandible, also showed a shorter base arch length, and less mandibular body length and ramus height, all of which leads to the smaller than average mandible.

Variable	JRA (Systemic) n=7 MEAN	SD	JRA (Poly) n=5 MEAN	SD	JRA (Pauci) n=3 MEAN	SD	Control MEAN	SD
Cranial								
S-N-Ar (°)	117.0	10.8	117.0	5.2	120.1	6.1	124.0	5.0
S-N-Ba (°)	132.5	6.9	127.9	4.7	130.7	7.4	130.5	5.0
Sagittal								
S-N-A (°)	80.9	4.9	84.6	3.4	83.5	9.0	81.5	3.5
S-N-B (°)	74.5	4.6	78.5	5.4	79.9	6.1	77.7	3.5
S-N-Pg (°)	73.8	4.8	78.5	5.7	79.3	6.7	79.0	3.5
A-N-B (°)	6.0	4.4	6.1	2.3	3.6	3.3	3.8	2.5
A-N-Pg (°)	7.4	5.2	6.1	2.8	4.2	3.3	2.5	2.5
ML/RLar (°)*	129.6	2.8	124.8	3.3	124.3	1.8	123.3	5.0
ML/MBLar (°)**	14.7	1.7	17.2	2.6	18.1	0.9	20.9	3.0
Dental								
Overjet (mm)	4.5	2.9	5.4	2.3	2.2	1.4	3.0	2.5
Overbite (mm)	0.2	2.2	2.1	2.0	1.9	2.0	2.5	2.0
U1/Palatal plane (°)	110.7	7.6	109.0	2.8	105.9	5.8	111.0	6.0
L1/Mn plane (°)	91.2	7.0	92.5	9.4	101.2	8.2	98.0	6.0
Interincisal angle(°)	121.7	14.1	131.3	12.6	121.4	10.3	128.0	6.0
Vertical								
Palatal plane/SN (°)	8.9	2.1	8.0	4.7	7.0	3.0	7.0	3.0
Mn plane/SN (°)	45.2	5.7	35.3	4.5	34.9	3.8	29.8	6.0
Palatal pl/Mn pl (°)	36.4	5.7	27.3	3.8	31.5	1.7	23.0	5.0

* Measures mandibular morphology using the ramus line to mandibular plane through Ar

** Measures mandibular morphology as the β angle

TABLE 1. Comparison of the cephalometric morphology of three different types of JRA patients and normal control subjects.

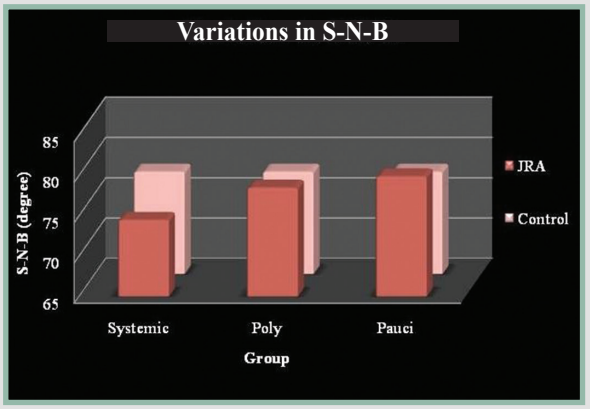

Figure 8. Variations in S-N-B in systemic, poly, and pauci JRA patients

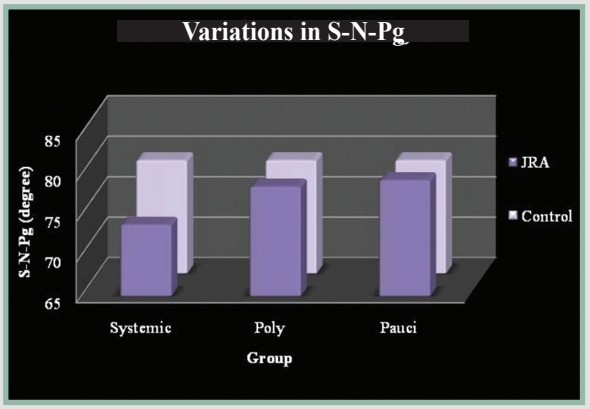

Figure 9. Variations in S-N-Pg in systemic, poly, and pauci JRA patients

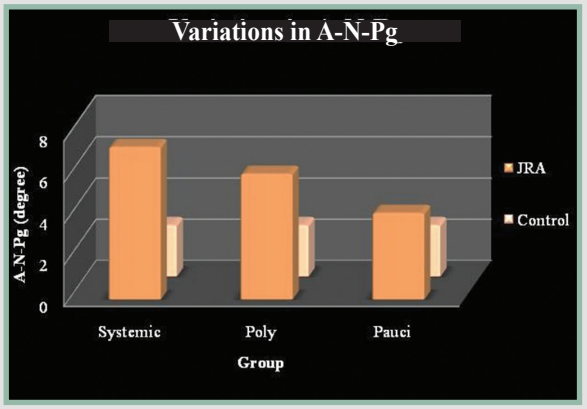

Figure 10. Variations in A-N-Pg in systemic, poly, and pauci JRA patients.

The angles Mn plane/SN (Figure 12) and palatal plane/Mn plane, representing the vertical skeletal dimensions, are significantly larger in all three groups, indicating a skeletal open bite and posteriorly inclined mandible.

The L1/Mn plane, the lower incisor inclination relative to the mandibular plane, is smaller—indicating that the lower incisors follow the mandibular rotation, contributing to an anterior open bite.

DISCUSSION

The most extreme craniofacial changes, particularly in the mandible, are found to be associated with the systemic type of the disease in this study.

Delayed skeletal maturation was observed in 33% of JRA patients (Figure 13), while 54% have similar chronological and skeletal age. Moreover, 13% of patients are found to be about one year ahead of their chronological age. The average period of delay is estimated to be two years.

This growth retardation is found to be associated with the systemic type of the disease. The duration and severity of disease, immobilization, poor nutrition and high doses of corticosteroids are considered to be the main factors that contribute

to growth impairment in these patients. However, the therapeutic regime of the JRA children was not taken into consideration in this part of the study and requires additional separate investigations.

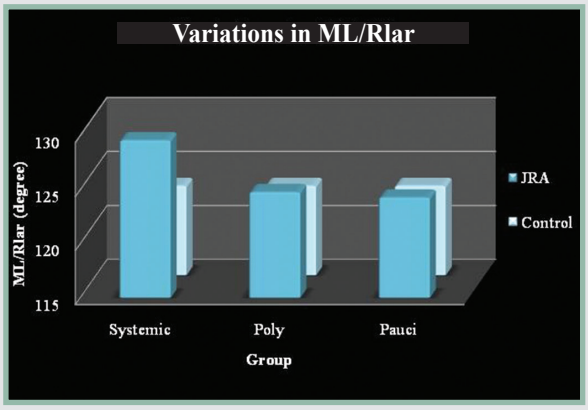

Figure 11. Variations in ML/Rlar in systemic, poly, and pauci JRA patients

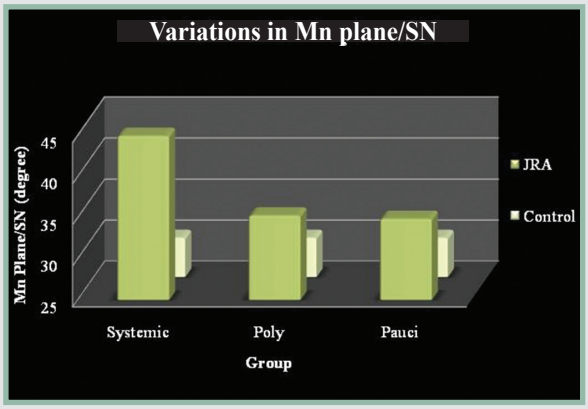

Figure 12. Variations in Mn plane/SN in systemic, poly, and pauci JRA patients

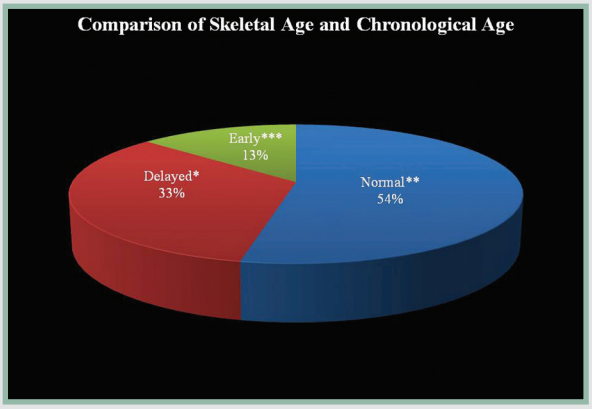

Figure 13. Comparison of skeletal age and chronological age of JRA patients

* Skeletal age is 2 years delayed from chronological age.
** Skeletal age is about the same as chronological age.
*** Skeletal age is 1 year earlier than chronological age.

CONCLUSION AND FUTURE WORK

This retrospective pilot study confirms earlier findings that systemic JRA patients have the typical facial characteristics associated with this disease. These features include a retrognathic mandible and posterior inclination of the mandible, both of which result from condylar inflammatory destruction of the TMJ. Early recognition in the daily clinical practice of these TMJ condylar changes is important in order to avoid further aggravating the developing JRA situation. This is particularly the case when a patient has an open bite that cannot be associated with airway problems or oral habits. Moreover, changes in condylar morphology may require further investigation using a CBCT scan if they are initially diagnosed on a Panorex. In some cases, it may also be necessary to use functional appliances to prevent the side effects of the disease on the occlusion.[9] In addition to conventional orthodontic treatment, TMJ or orthognathic surgery should be considered in severe cases, but generally not until the disease has burned out. Future studies should include more JRA patients than available for this pilot study so that a reliable statistical analysis can be performed. It is also recommended to include hand-wrist radiographs to assess patients' skeletal age in order to determine whether skeletal maturation is delayed as a result of medical treatment, or the disease itself.

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—W.W.T.