Craniofacial growth in juvenile chronic arthritis

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The craniofacial growth in children with juvenile chronic arthritis (JCA), especially that of the mandible, and the degree of destruction of the mandibular condyles vary depending on the heterogeneity in duration and intensity of the disease. In JCA children showing destruction of the temporomandibular joint, the dentofacial morphology is characterized by overall smaller dimensions of the mandible, mandibular retrognathia, a steep mandibular plane, Class II malocclusion, dental crowding, and frontal open bite. In children with unilateral condylar destruction, asymmetries will develop, with the chin deviating to the affected side. The facial morphology of JCA children with condylar lesions becomes more abnormal during growth, reflecting a decelerated mandibular development and a backward-rotating growth pattern. The main single cause of the deviating craniofacial growth is mandibular condylar destruction. Other factors that may influence the craniofacial growth are head posture, soft tissue stretching, disease activity and drug therapy, type of onset of the disease, muscle weakness, decreased functional ability, and orthodontic treatment. □ *Children; facial morphology; juvenile rheumatoid arthritis; orthodontic treatment; Still's disease*

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Juvenile chronic arthritis (JCA) is characterized by chronic inflammation in one or more joints. The course of the disease is unpredictable and the etiology remains unknown, though current opinion favors the hypothesis that immunologic mechanisms play a major role (1). Precipitant factors such as trauma, heredity, and infections are discussed in the development of the disease.

The prevalence in child populations, using the criteria of the European League Against Rheumatism (EULAR) (2), has a range of 60–80 per 100,000 children. The annual incidence is about 11 per 100,000 children, with girls predominating over boys at a ratio of about 3:2 (3). About 50% of the children show remission of the disease after 5–10 years (3). The prognosis for JCA is most favorable among those children with a pauciarticular form of the disease.

There is no uniform, worldwide agreement on the use of diagnostic criteria for chronic arthritis in children. The criteria and nomenclatures most frequently applied at present are the North American criteria, according to which the disease is named juvenile rheumatoid arthritis (JRA) (4); the EULAR criteria, using the name juvenile chronic arthritis (JCA) (2); and the English criteria, using the name Still's disease or juvenile chronic polyarthritis (JCP) (5).

Temporomandibular joint

The most important growth center of the mandible, especially early in life, is located in the condylar joint. Early arthritic destructions of the chondral part of the condyle may subsequently seriously affect mandibular development and growth.

The most common signs of arthritis in the temporomandibular joint (TMJ), caused by the inflammation in the synovial membrane, are erosion and flattening of the condyle (from small bony erosions to complete absence of the condylar head) (6–8). Erosive changes, flattening and sclerosis of the temporomandibular fossa, narrowing of the joint space, anterior displacement of the condyle in the fossa, osteophyte formation, subchondral cysts, reduced condylar size, and restricted translatory movement of the condyle are other signs found (7, 9–13).

In studies using the magnetic resonance imaging technique (MRI), it is possible to visualize details of soft tissue anatomy including cartilage, ligaments, muscles, and other soft tissue structures. Using MRI Taylor et al. (14) found cortical erosion, thinning, and perforation of the disk, reduction of joint movement, locking, pannus (a hyperplastic and granulomatous synovial membrane that grows to extend over the articular surface), and effusion (escape of fluid into a tissue) among 15 children with JRA.

Asymmetry between the two condyles is also a common finding, seen especially at the onset of the disease (7, 8, 15, 16).

The lesions described above are found in up to 63% of children with chronic arthritis (6–8, 11, 17–20) when diagnosed with conventional radiography or computer tomography. A higher prevalence is found among children with polyarticular onset as compared with pauciarticular or systemic onset (21–23). The considerable differences in prevalence result from the fact that the observations were obtained from different clinical materials (variation in age of the patients and duration of the disease; different diagnostic criteria; population-based or referred subjects, etc.) and from different types of radiographic investigations. Studies using MRI for diagnosing arthritic changes

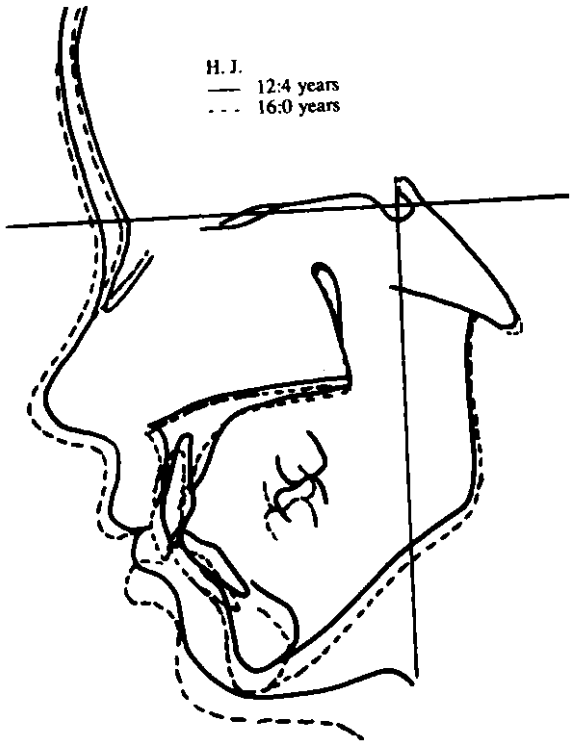

Fig. 1. Craniofacial growth in a girl with juvenile chronic arthritis without visible condylar lesions. A normal growth pattern was seen during the period studied.

such as cartilage damage and erosions have shown increased sensitivity compared with those using conventional radiographic techniques (24), indicating that the percentage of TMJ arthritis in JCA children is probably underestimated.

Craniofacial morphology and growth

Children without condylar lesions

In JCA children without radiographically visible condylar lesions, a mainly normal facial morphology is observed, in contrast to those with lesions (25, 26). The growth changes are similar to those in healthy children with normal occlusion (Fig. 1). However, a tendency toward a steeper mandibular plane, an increase in anterior facial height, and smaller mandibular dimensions have been observed even among these JCA children (15, 27).

Furthermore, it is likely that a considerable percentage of children with chronic arthritis will develop condylar destructions during their growth period (20), especially children with a progressive disease with severe attacks and those who develop a polyarticular course of the disease (21). These children will have an increased risk of developing an abnormal dentofacial growth pattern.

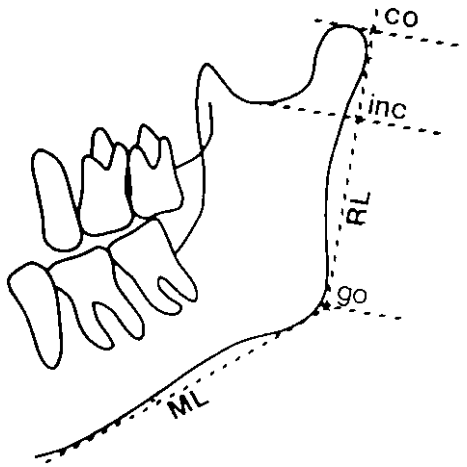

Fig. 2. The condylar ratio: the distance 'co–inc' divided by the distance 'inc–go', expressed as a percentage. RL = tangent to ramus mandibulae; ML = tangent to lower border of mandible.

Children with condylar lesions

In JCA children showing destructions of the TMJ, the dentofacial morphology deviates from that of normal children, especially with regard to mandibular morphology and position, with overall smaller dimensions of the mandible, mandibular retrognathia, a steep mandibular plane, and anterior apposition of bone at the chin point (25, 28–31). The anterior alveolar height is found to be large, and the posterior lower facial height is small (25, 28–30). The gonial angle is obtuse, and there is a more pronounced antegonial notching (9, 28, 31). The condyle of JCA children is positioned more anteriorly in the fossa (7, 32).

The relation between the condylar and ramus height, expressed as the condylar ratio (co–inc/inc–go, %) (Fig. 2), is significantly smaller in a JCA group of children than in two healthy groups of children with either Class I or Class II malocclusion (16).

In most studies the maxilla is found to be smaller in the vertical dimensions and is also posteriorly rotated in affected children. The findings may be explained as a secondary reaction to the decreased growth of the mandible or to an altered loading of the posterior regions of the maxilla (26, 28, 31–33).

In children with unilateral condylar destruction, asymmetries will develop, with the chin deviating to the affected side. The gonion point and the maxillary basis are placed more cranially, and the alveolar process seems to be compressed, resulting in shorter vertical dimensions on the affected side (34, 35).

Growth changes

The facial morphology of the JCA children with condylar lesions becomes more abnormal during growth,

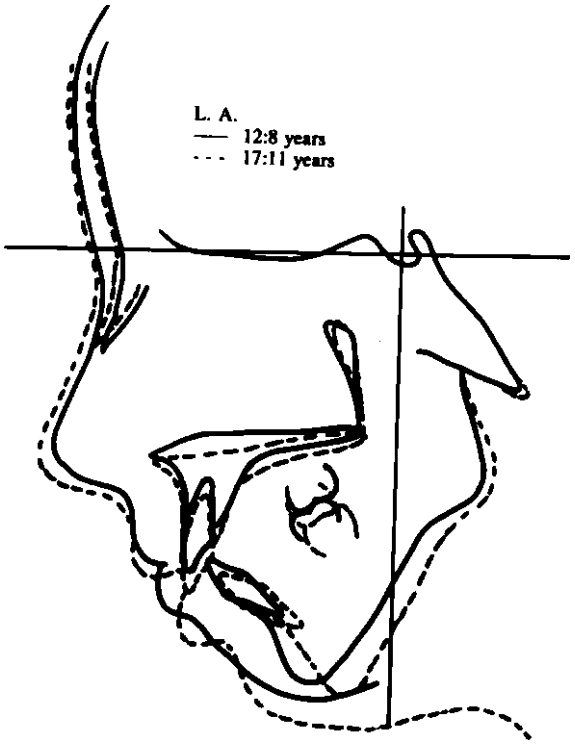

Fig. 3. Craniofacial growth in a girl with juvenile chronic arthritis with severely destroyed mandibular condyles. She exhibited a backward-rotating growth pattern and abnormal, decelerated mandibular development.

reflecting a decelerated mandibular development and a backward-rotating growth pattern (Fig. 3) (27, 31, 32, 36).

Signs of absolute shortening of the ramus height and the total mandibular length during growth have also been observed (27, 37). However, spontaneous restitution with new bone formation might increase the ramus height during periods of low disease activity or as a result of successful medicamental treatment (38).

Björk & Skieller (31) studied the posterior rotation of the mandible using implants in one girl with JRA. Collapse of the condylar structures caused the posterior part of the mandible to move upward and the anterior part downward. The center of rotation was located at the occlusal level of the molars, which were lowered less than normally.

This rotational pattern observed in the child with JRA differed from that observed in connection with muscular dystrophy (39) or in long-face syndrome (40). In these cases a lowering of the maxillary molars exceeded the vertical growth component of the condyles, which also causes a posterior rotation of the mandible and a downward growth of the symphysis.

Occlusion

Class II malocclusion is found to be almost twice (about 30%–40%) as common in children with chronic arthritis as in healthy children (9, 11, 17, 25). Reduced overbite and frontal open bite (6, 8, 9, 17, 25, 29) have also been found. Crowding, both general and local, especially in the lower incisor region, has been described (25, 31). JCA children show significantly more proclination of the lower incisors. During growth a decrease in overbite and an increased frontal crowding in the lower jaw as well as changes from a Class I to a Class II molar relation are observed (27).

Factors influencing the craniofacial morphology and growth

Condylar lesions

Radiographically visible condylar lesions, as a result of the inflammatory influences of the joint structures, seem to be the most important single factor influencing the mandibular growth, and thus the facial morphology and occlusion, of children with chronic arthritis (12, 15, 17, 30, 31, 41, 42). A higher degree of mandibular retroposition and smaller mandibular dimensions are found in patients with complete destruction of the head of the condyle compared with those with partial destructions (15, 25, 29, 42).

Head posture

Solow & Kreiborg (43) suggested that a lack of forward growth of the mandible initiates an increased extension of the head in relation to the cervical column in order to maintain an adequate airway. This may result in stretching of the soft tissue, which has a restraining effect on the facial development. It may also be a reason for gonial notching.

Disease activity and drug therapy

Early onset, long duration, and aggressiveness of the disease and long-standing corticosteroid therapy are correlated with TMJ abnormalities and consequently with reduced growth of the mandible (11, 15, 18, 21, 29, 41).

Poly-, pauciarticular, or systemic onset of disease

Polyarticular onset affects the facial morphology more severely than pauciarticular or systemic onset (18, 21, 22).

Mericle et al. (22) found different effects on facial growth depending on whether the onset of the disease was pauci- or polyarticular. They stated that the facial pattern in pauciarticular onset was superficially similar to long-face syndrome, with an increased posterior facial height and mandibular length and a retrognathic mandible. They suggested that the altered masticatory function, owing to a lower degree of condylar loading, may be responsible for this pattern. In polyarticular onset cases, the disease results in more severe damage to the condyles and should thus affect the face and mandible differently, resulting in a

smaller, more retrognathic, and more steeply angulated mandible (i.e. as described above).

In systemic onset JRA, Hanna et al. (23) found only minimal effects on the facial morphology compared with that of controls, such as a mild downward and backward mandibular rotation and a more convex profile. The findings were strongly correlated to the children with evident condylar changes.

Stabrun et al. (15) even found reduced mandibular growth in affected children without visible condylar lesions. They found that long duration of the disease and the type of disease at onset were associated with smaller mandibular dimensions despite the absence of visible condylar lesions. It is likely that early inflammatory changes, undiagnosed with conventional radiography, have occurred in the TMJ of these children (44).

Muscular weakness

Muscular weakness in correlation with dentofacial morphology and occlusion has been thoroughly investigated in several clinical studies of non-rheumatic subjects, where it was found that those with long facial proportions exert lower forces than 'normal' or 'short-face individuals' (45–48).

Maximal molar bite force in JCA children was found to be about 60% of that recorded in healthy children (49). In a group of adults with rheumatoid arthritis (50), this force was about 20%–25% of that of the healthy controls. The low forces recorded among subjects with arthritis might be caused by inhibition due to pain or muscular atrophy, in turn due to inactivity and disuse of the musculoskeletal system (51). It might, however, also be the result of a type II fiber atrophy, due to direct involvement of muscle fibers in the inflammatory process (52, 53). Wróblewski & Nordemar (53) stated that there seemed to be a direct correlation between fine structural changes and the intensity and duration of the disease. This can explain the remarkably low bite forces in the adult rheumatic group described above and may indicate that the low bite force in JCA children will decrease further with time.

It is reasonable to believe that the low muscular forces in JCA children might influence the craniofacial morphology during growth, although so far no study has proven any significant correlations between low muscle forces and morphology in this group of children (49, 54). The reason may be that an impairment of function may have occurred in connection with the active inflammatory phase of the disease, thereby delaying the influence on the facial morphology.

Jaw exercises

A weak masticatory musculature may be strengthened by jaw exercises (55), and jaw exercises were also found to have a positive influence on dentofacial growth in healthy children with a 'high-angle face' and frontal open bite (56, 57). Nordemar et al. (58) found that exercises might reduce the destruction of joints other than the TMJ. They found that physical training over 4–8 years resulted in retardation of the destruction of the arthritic joints of the lower extremities in patients with rheumatoid arthritis.

Orthodontic treatment

The high prevalence of malocclusions among JCA children increases the demand for orthodontic treatment. In most JCA children orthodontic treatment can be carried out satisfactorily during growth by using both functional and fixed appliances (27). However, one might expect less response to treatment with functional appliances in children with chronic arthritis owing to the condylar lesions, especially in those children who are in poor general health and/or develop more severe condylar lesions during the orthodontic treatment period. On the other hand, in some JCA children good treatment results may be achieved despite extensive condylar lesions. This may be explained by the different rates at which the condylar lesions progress. In those children with good treatment response, the lesions may have progressed slowly, or new hard tissue may even have formed during periods of low disease activity. The compensatory mechanisms during growth may also have had 'sufficient time' to counteract the negative effects of the lesions. In contrast, continuous and rapid destruction may result in poor treatment effects and may also be one reason for the relapse seen in some JCA children.

In conclusion, successful management of disturbances in the facial region requires a good knowledge of dentofacial growth and the function of the stomatognathic system. It is recommended that all children with a diagnosis of JCA should be followed up as regards dentofacial morphology and signs and symptoms of TMJ arthritis. This is important because of the high prevalence of involvement of the TMJ in the disease and the difficulty in predicting which children will show involvement. If treatment is started early during growth, it might be possible to counteract growth disturbances, to maintain mobility in the joints, and to maintain proper occlusion, thereby optimizing the local environmental conditions throughout the growth period.

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