Introduction

Juvenile idiopathic arthritis is a form of arthritis of unknown aetiology that appears before the 16th year of age and persists for at least 6 weeks [Petty et al., 2004]. The term arthritis means swelling or joint effusion or the presence of at least 2 symptoms among limitation of movement, pain during movement or pressure and temperature change to tactile stimulation [Fantini, 2000]. In the past it was known as “Juvenile Chronic Arthritis” in Europe and “Juvenile Rheumatoid Arthritis” in the United States of America [Petty et al., 1998].

Juvenile Idiopathic Arthritis is a collection of different diseases. According to the Edmonton’s classification there are different subtypes, each with specific clinical features:
- systemic arthritis;
- oligoarthritis;
- polyarthritis (negative for rheumatoid factor);
- polyarthritis (positive for rheumatoid factor);
- psoriatic arthritis;
- enthesitis-related arthritis;
- undifferentiated arthritis [Petty et al., 2004].

The prevalence of Juvenile Idiopathic Arthritis ranges from 0.07 to 4.01 every 1,000 children, while the annual incidence ranges from 0.008 and 0.226 every 1,000 children [Manners and Bower, 2002]. In the United States the juvenile arthritis affects 294,000 children [Helmick et al., 2008]. The prevalence of JIA in Catalonia was 39.7/100,000 [Modesto et al., 2010], while the prevalence in Western France was 15.7/100,000 children [Solau-Gervais et al., 2010].

The main signs of temporomandibular joint involvement are chronic synovitis with epithelial proliferation, increase in synovial fluid and inflammatory pressure [Ganik and Williams, 1986]. The condyle damage is variable, ranging from minimal erosion of the head to complete condylar decapitation [Walton et al., 2000].

Radiographically early condylar damage presents an initial step with soft tissues oedema and enlargement of articular space. A major involvement presents growing cartilage destruction and cortical bone erosion [Walton et al., 2000]. MRI exam is the gold standard for detection of the TMJ disorder, particularly for detection of early damages [Weiss et al., 2008].

The late phases of TMJ involvement exhibit high condylar resorption, condylar destruction, and flattened articular eminence.

At this stage the clinical signs could be reduced opening movement, articular sounds during movement, muscular and articular oedema, together with pain at rest and during movement, morning rigidity [Bellintani et al., 2002].

Most of these children have no symptoms and clinical findings of joint temporomandibular, so it is very difficult to assess TMJ involvement [Pedersen et al., 2001].

The condyle is an important site of growth and therefore its involvement has important consequences on the lower third of the face [Pruzansky, 1973; Ringold et al., 2009; Kreiborg et al., 1990]. In the condyle there are severe alterations with its displacement and total cartilage destruction, meanwhile there are no pathological changes in the temporal fossa [Pirttiniemi et al., 2009]. Endochondral mandibular growth can be disturbed when the condyle is being resorbed or eroded [Hu et al., 2009].

In the case of bilateral involvement an important mandibular hypoplasia with mandibular retrusion can be observed, and a characteristic facies, named bird face, is observed [Ronning et al., 1994].

If only one condyle is involved, marked asymmetry and lateral deviation to the affected side during opening can be observed. These signs are connected to the asymmetric growth of the two condyles, where the involved side presents growth deficits [Corradi et al., 1994; Pedersen et al., 2001].

Typical morphological alterations, such as mandibular
hypoplasia, are also related to the corticosteroid therapy prescribed to these patients [Kjellberg et al., 1995]. In a study on animal models the treatment with corticosteroids may result in even more pronounced mandibular growth reduction than that caused by arthritis alone [Stoustrup et al., 2008]. Another study, on the contrary, is in favour of corticosteroids therapy, which according to the author could increase mouth opening [Ringold et al., 2008].

In addition to mandibular hypoplasia, also ramus height and posterior vertical dimension reductions, with subsequent discrepancies between anterior and posterior facial dimensions, can be observed. [Kjellberg et al., 1995]. These alterations result in clockwise mandibular growth [Gianni et al., 1987]. There is also a homeostatic reaction of the body with bone apposition in the gonial region and antegonial notching.

Several studies tried to illustrate the cephalometric features of these subjects [Kjellberg et al., 1995; Ronning et al., 1994; Billiau et al., 2007; Sidirooulou-Chatziagianii et al., 2001]. Many of them present skeletal Class II, wide gonial angle, clockwise growth of the mandible and reduction in the posterior vertical facial height [Kjellberg et al., 1995; Gianni et al., 1987; Ronning et al., 1994].

Head position is usually extended in order to permit normal breathing even if a mandibular retrusion is present [Ronning et al., 1994].

**Treatment protocol**

Collection of medical history includes the following:
- age of onset of the JIA;
- disease subtype;
- sex and family history;
- age at onset of [TMJ] involvement (defined as first complaints of the patient and/or radiographic alterations);
- unilateral or bilateral temporomandibular involvement;
- symptoms linked to temporomandibular involvement (headache, grinding, crepitations, clicking, reduced mouth opening, inability to eat hard food).

Clinical examination includes facial considerations, intraoral examination and functional examination of the [TMJ].

Functional examination of temporomandibular joint focuses on condylar movement during opening and closing, the presence of mandibular deviation during opening, maximum vertical opening.

Manual examination of the condyle helps to detect abnormal condylar movements. Auscultation of the [TMJ] using a stethoscope is also performed. Clinical examination includes electromyographic and kinesiographic tests.

Treatment entails placement of an activator fabricated through a wax bite registration: a piece of warmed wax is placed on the upper dental arch and the patient is guided to close the mouth in mandibular protrusion with coincidence of upper and lower midlines [Bondi, 1994]. The registration bite might be very thin in order to obtain minimum vertical dimension. The resulting activator is made of resin, with a central screw and a vestibular arch.

The functional treatment is recommended before the growth pattern becomes pathological [Pedersen et al., 1995]. However, recently some authors reported that JIA progression makes the mandibular growth more favourable, while the condylar damage persists [Fjeld et al., 2010; Arvidsson et al., 2010].

In our opinion the functional treatment protects the joint, avoiding the destructive action of synovitis on the temporomandibular region. At the same time, the appliance has an orthopaedic function and modifies the unfavourable growth of the mandible helping its counter-clockwise rotation [Farronato et al., 2009].

The appliance is modified every two months increasing its posterior vertical dimension in order to stimulate growth of the mandibular ramus because of the dislocation of the mandibular condyle. The activator helps the growth of the mandibular ramus with increase in posterior vertical facial height and counter-clockwise rotation with subsequent improvement of the occlusion, masticatory function and profile [Bellintani et al., 2005].

**Case report**

A 8-year-old male was referred to our department by his rheumatologist, who had made the diagnosis of Juvenile Idiopathic Arthritis 1 year earlier and had treated it with [NSAIDs].

The clinical examination showed remarkable asymmetry of the left mandible with reduced left facial height.

The intraoral examination underscored occlusal plane inclination and noncoincidence of the upper and lower midlines (Fig. 1-5).

Mandibular kinesiology presented a reduced opening movement and deviation to the affected side during opening, in addition to articular sounds in the left side.

Frontal and lateral radiographs, panoramic radiograph and tomographic images were also analysed (Fig. 6-8): the left mandibular ramus was shorter than the right one and the left condyle was flattened. Lateral cephalometry showed skeletal Class II with mandibular retrusion and clockwise rotation, hyperdivergence and increased anterior vertical facial height. The frontal cephalometric analysis confirmed the marked asymmetry detected during the clinical examination. The tomographic images showed condylar derangement and a reduced translation of the condyle during opening.
The patient underwent functional treatment using the activator described in the protocol with midlines coincidence. The affected side was periodically increased in height, while the healthy side was not modified.

At the end of the treatment facial and intraoral asymmetry were improved and the articular sounds disappeared (Fig. 9-15). Clinical and kinesiographic examinations showed improvement in mandibular opening.

The follow-up up 5 years after the end of therapy, showed results stability and improvement in mandibular kinesiology.
Conclusion

In this case report, the treatment had several advantages, with improvement of the clinical signs of the disease, and permitted to obtain their stability over time.

The case reported underscores the efficacy of the treatment proposed also for a case of unilateral temporomandibular joint involvement.

References

Arvidsson LZ, Fjeld MG, Smith HJ, Flata B, Ogaard B, Larheim TA. Craniofacial growth disturbance is related to temporomandibular joint abnormality in patients with juvenile idiopathic arthritis, but normal facial profile was also found at the 27-year follow-up. Scand J Rheumatol 2010;39(5):373-9.


