

Long term results of idiopathic hemifacial palsy: Orthodontic and surgical multidisciplinary management



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ABSTRACT

Idiopathic facial palsy is the most common cause of unilateral neurologic disorders of the cranial nerves.

It can result from congenital, due to delivery traumas, and genetic or malformative diseases, or acquired due to infective, inflammatory, neoplastic, traumatic or iatrogenic causes. In the great majority of cases its causes are unknown.

A careful differential diagnosis is recommended in paediatric patients, in order to establish the most appropriate treatment as the therapeutic approach differs in relation to the aetiology. Controversy exists regarding treatment options. The primary concern in the treatment is to restore the aesthetics, function and comfort. Facial palsy may result in a multitude of conditions such as: affected salivation, taste and lachrymation, depending on the topography of the facial nerve affection, and patients may also refer auditory hyper-sensitivity which may result in noise intolerance.

In this paper a case of a child with unilateral facial palsy is described. The patient was treated by orthopaedic appliances and surgical facial reanimation. At the end of therapy good aesthetic and functional results were obtained. Oral functions as well as facial expression were partially recovered and symmetry was almost completely restored.

Children with idiopathic facial palsy have a better prognosis if the diagnosis is early and treatment is multidisciplinary.

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1. Introduction

Idiopathic facial palsy is a medical condition, with an incidence of 20–25 cases per 100,000 individuals per year [1–4] with a peak usually between the age of 15 and 50 years [5–7]. It is rare in children younger than 10 years old [5]. Ozkale and Barr [8–10] reported an estimated incidence of about 6.1 cases per year per 100,000 in children aged between 1 and 15 years.

There do not seem to be gender differences [11,12] and no predilection concerning face side.

In fact, if not treated, it can cause functional and aesthetic outcomes.

The pathogenesis remains controversial. It can either be congenital due to delivery traumas or genetic or malformative diseases, or acquired due to infective, inflammatory, neoplastic, traumatic or iatrogenic causes.

In approximately 40–75% of the cases, the cause still remains idiopathic [8].

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Idiopathic facial palsy is commonly known as Bell's palsy as it was described by Dr. Charles Bell in 1821 as an acute peripheral facial nerve palsy usually of unknown cause [13]. It is typically unilateral and can be complete or partial [13,14]. It represents a clinical problem with several implications, particularly when occurring in childhood. The condition leads to the



Fig. 1. Extra-oral view: soft tissues asymmetry, right mandibular deviation.



Fig. 2. Extra-oral view: inability to close the right eye and to corrugate the right eyebrow, absence of right naso-labial groove mobility.

partial or complete inability to voluntarily move facial muscles on the affected side of the face, and can exacerbate important asymmetries of the face.



Fig. 3. Intra-oral view: deciduous dentition, Class III on the right side, Class I on the left side, deviation of the midlines.

Knowledge and function of the VII facial nerve is crucial to understand the pathophysiology of Bell's palsy [5]. The VII cranial nerve is embryologically derived from the second branchial arch. It consequently innervates all structures originating from it [15].

The nerve provides efferent motor innervation to the muscles of the face, the stapedius, and the posterior belly of the digastric muscles. Also sensory and parasympathetic fibres travel with the facial nerve. These fibres supply the lacrimal and the submandibular glands [5]. Therefore, the facial nerve not only provides conscious and subconscious motor control of the facial musculature, but also enables speech articulation [15]. Additionally, it facilitates the ability to smile. Consequently, facial nerve injury may inflict a spectrum of functional aesthetic and psychosocial insults. Stapedial reflex may be absent. Studies report cases of patients with Bell's palsy who reported hearing loss and they agree that the cause of hearing loss in these patients can be the absence of stapedial reflex. The reflex is derived from bilateral contraction of stapedial muscle in the middle ear, in response to loud sounds, which occurs at about 85 dB HL. The patient reported in this study didn't present any of the symptoms described.

Affected patients are usually unable to close their eyes, leading to potential eye injury, saliva dribbles down the angle of the mouth, facial appearance becomes asymmetric [13].

Some patients may complain of noise intolerance or lost of taste sensation [13], speech and language involvement.

Long term poor outcomes can be devastating to the patient.

A careful diagnosis is recommended in paediatric patients in order to establish the most appropriate treatment.

Treatment is controversial. It is generally designed to improve facial function.

Patients should ideally be assessed and managed by a multidisciplinary team involving paediatrician, orthodontists, maxillo-facial and plastic surgeons, psychologist, speech and physiotherapist, neurologist and otolaryngologist.

Most patients with Bell's palsy recover without any cosmetically clear deformity. 30% of patients, however, experience long-term symptoms following the paralysis, and approximately 5% present a high degree of sequelae [16]. Bell's palsy sequelae include incomplete motor or sensory regeneration and aberrant reinnervation of the facial nerve.

The aim of this paper is to describe a clinical case of a paediatric patient presenting a congenital facial palsy. The therapeutic strategies consisted in surgical techniques associated to rehabilitative orthodontic approaches. The case described

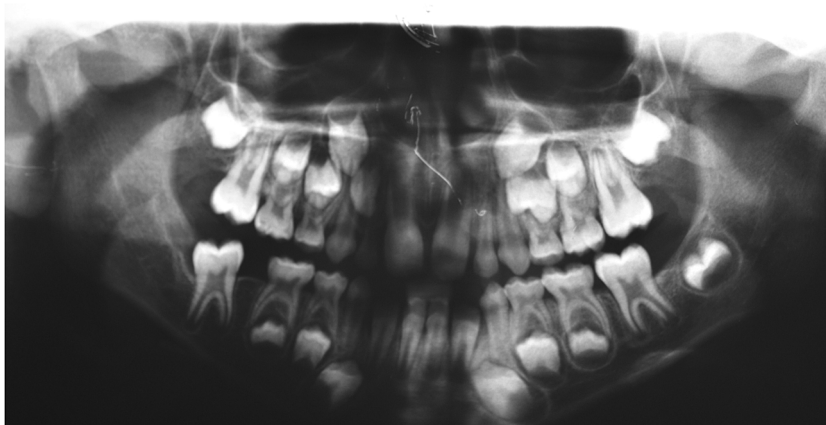


Fig. 4. Panoramic film shows left lateral incisor, right second and third permanent molars congenitally missing.

emphasizes how idiopathic facial palsy may be compensated by the correct growth of the muscles and skeletal facial structures. The treatment can lead to the best possible aesthetic and functional results.

2. Case report

A 5 years and 11 months patient was referred to the orthodontic department of Milan University by his paediatrician. He had no previous significant medical history except for a diagnosis of hemifacial Bell's palsy of unknown origin as biochemical and imaging investigations carried out didn't identify any evident cause.

Parents denied any type of facial trauma or systemic affliction. Their major concern was unaesthetic asymmetry in common facial mimic. They referred that since he was newborn he showed an asymmetry of the face with buccal deviation when crying and that the lacrimal and salivary production was reduced.

Clinical examination revealed a slight asymmetry of the soft tissues, a lateral deviation of the mandible to the right side, an inability to close the right eye and to corrugate the right eyebrow, restricted movement of both the superior and inferior lips, absence of naso-labial groove mobility. (Figs. 1 and 2).

Intraoral examination revealed deciduous dentition, Class III on the right side, Class I on the left side, deviation of the midlines (Fig. 3).

Upper maxillary arch was constricted and hypoplastic.

Panoramic examination showed left lateral incisor, right second and third permanent molars congenitally missing (Fig. 4).

Lateral headfilm showed a skeletal Class III with high anterior vertical dimension (Fig. 5).

Frontal headfilm confirmed the deviation of the mandible to the right side (Fig. 6).

The nerve palsy was classified between grade 3 and 4 according to the House-Brackmann facial nerve grading system [17]. Percentage of success in those cases is highly affected by the paralysis degree according to House-Brackmann scale: patients degree of paralysis during the first visit, before starting myofunctional physiotherapeutic exercises, was IV grade.

After one month since the first visit in 2001 it was decided to expand the maxillary upper arch with a Hyrax rapid palatal expander to correct the crossbite and to favour the correct position of the mandible. The expander was activated twice a day until the desired expansion was obtained. Then it was left in place for six months as retention. Meanwhile the patient and the parents were advised and instructed to start facial physiotherapy myofunctional exercises and to keep the right eye close with tape or a night mask during the night to avoid conjunctive dryness.

After six months of retention the rapid palatal expander was removed and multibracket were placed. to solve the cross bite.

After 6 years in 2007 the patient was referred to the surgeon for facial reanimation in order to undergo surgical treatment. The microsurgical technique consisted in a two stage operation: The first stage consists in sural nerve grafts between the branches of the facial nerve of the healthy side of the face and those of the injured side (cross facial nerve grafting). Branches of the non-paralyzed facial nerve are exposed and used as donors to motor the cross facial nerve graft. The graft is micro-surgically repaired to distal functioning nerve branches, allowing re population of the nerve graft with sprouting axons used to innervate a functional muscle in 6 months to a year after the first stage operation.

The second stage consists in functional muscle transplantation where simultaneous gracilis muscle harvest and preparation of the paralyzed side for the face are performed by two microsurgical teams.

This practice allows the healthy facial nerve to send a symmetrical and synchronous pulse to the paralyzed side [8,18–23].

After surgery, in 2008, a removable functional appliance characterized by the presence of a lateral resin shield on the side of the hypoplasia was applied. This treatment was conducted in order to gain a physiological growth of both the jaws and soft tissues. The aim of this appliance was also to promote the growth of the right side of the mandible and to correct the asymmetry.

Facial expression exercises, as myofunctional physiological therapy, were also recommended in order to reduce muscles stiffness and facilitate facial movements.

By the end of 2009 fixed orthodontic appliance was applied in order to create space between upper left central incisor and canine for implant rehabilitation.

At the end of therapy, in 2010, the appliance was removed and an essix retainer was delivered to prevent undesired dental movement and to prevent extrusion of the 17 before the placement of an implant and a Maryland prosthetic bridge was placed to substitute the 22. by the end of 2014 18 was extracted as it presented no antagonist.

The facial examination showed a better symmetry of the face (Fig. 7). The intraoral examination permitted to observe a dental Class I on both sides, midlines coincidence, overbite and overjet within the norm (Fig. 8).

Panoramic film showed good symmetry of the roots (Fig. 9).

Lateral headfilm showed a skeletal Class I and correct vertical dimension (Fig. 10).

Frontal headfilm showed no asymmetries (Fig. 11).

A Maryland bridge was applied to substitute the left lateral incisor while waiting for implant placement.

After treatment the patient recovered normal facial function and after 1 year of follow-up, facial movements were satisfactory. No signs of recurrence were noted.

Follow-up checkups were programmed.

Although a slight asymmetry of the soft tissue on the side of palsy was still present, the face of the patient was aesthetically satisfactory and the patient and his parents were very satisfied with the obtained results.

Ten years follow up examination confirmed unlimited and pain free functioning of masticatory muscles with excellent performance. No disturbance of mandibular or facial growth was evident as the patient was followed through the growth period.

Stable relationship with good alignment of dental arches and correct overjet and overbite were maintained (Fig. 9).



Fig. 5. Lateral headfilm shows a skeletal Class III with high anterior vertical dimension.



Fig. 6. Frontal headfilm confirms the deviation of the mandible to the right side.



Fig. 7. Extra-oral view: better facial symmetry at the end of treatment.

Radiographic examinations confirmed the stability of the results (Figs. 9–13) (see Figs. 14–16).

3. Discussion

This report wants to propose a novel treatment procedure including surgical, myo-functional, orthodontic and prosthetic long term approach to a very complicate case presenting a multitude of problems which cannot be standardized and extended



Fig. 8. Intra-oral view: dental Class I on both sides, midlines coincidence, overbite and overjet within the norm; a Maryland bridge is applied to substitute the left lateral incisor while waiting for implant placement.



Fig. 9. Extra-oral view: facial symmetry ten years follow up.

to other cases due to the singularity of the treated patient. Several pathologies can be included in the differential diagnosis of Bell's palsy. Diagnosis depends on clinical signs, symptoms and laboratory investigations to exclude other possible causes of



Fig. 10. Intra-oral view: ten years follow-up.

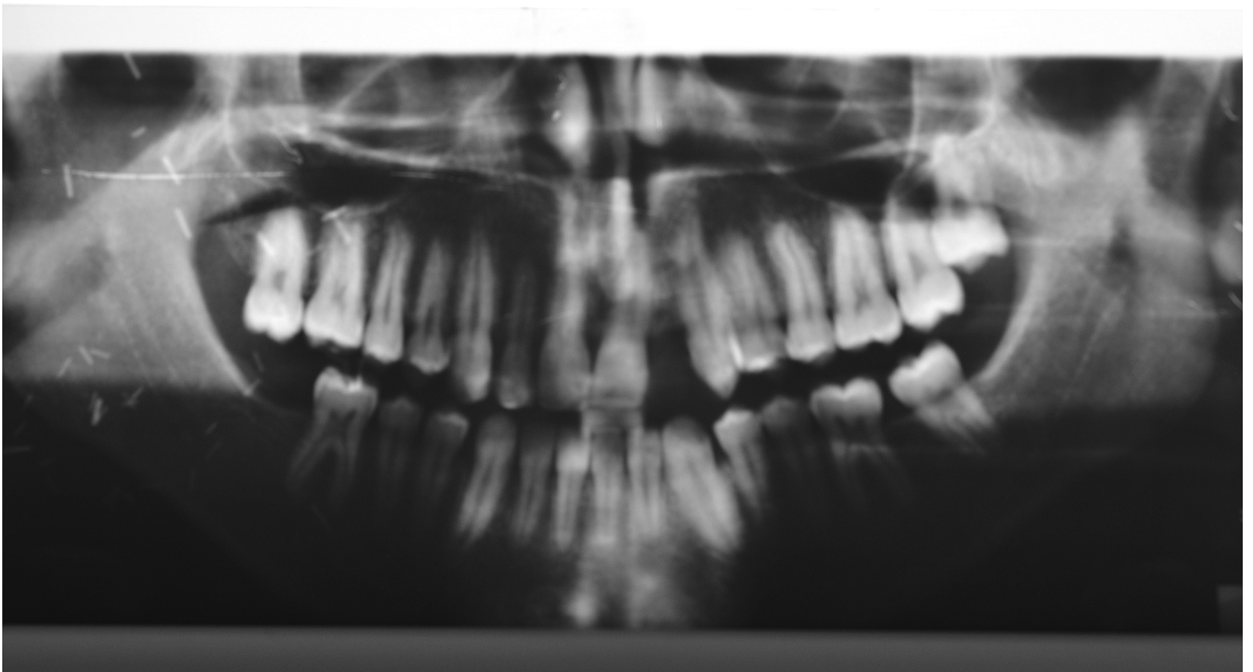


Fig. 11. Panoramic film shows good symmetry of the roots at ten years follow up.

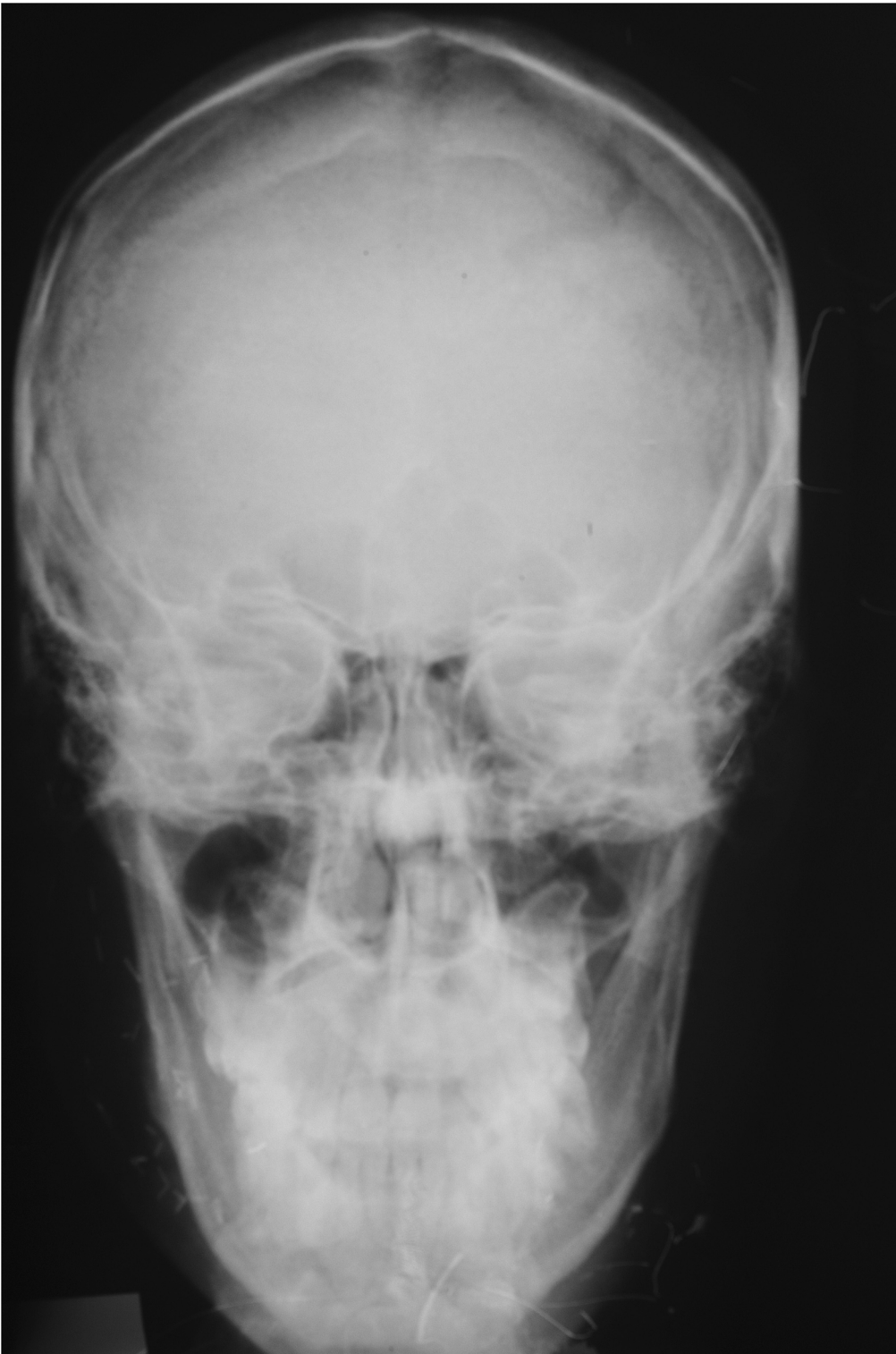


Fig. 12. Lateral headfilm shows a skeletal Class I and correct vertical dimension at ten years follow-up.

palsy. Idiopathic facial palsy, is the most common mono-neuropathy, or disorder affecting a single nerve, and is the most common diagnosis associated with facial nerve weakness/paralysis.



Fig. 13. Frontal headfilm shows no asymmetries at ten years follow-up.

This condition leads to the partial or complete inability to voluntarily move facial muscles on the affected side of the face. It represents a serious clinical problem as it can cause functional and aesthetic outcomes, above all in growing patients. The most alarming symptom of Bell's palsy is the paralysis of muscles of facial expression and often causes significant facial distortion.



Fig. 14. Shows three years post op. in animation.

The degree of paralysis represents a prognostic element. Patients with partial paralysis have a better prognosis [8]. According to House-Brackmann scale the II degree has a good outcome, while the III and IV degrees are associated to moderate residual dysfunctions. The V and VI degrees, instead, have poor possibility of recovery [5,8,24]. 3***.



Fig. 15. Shows three years post op. in animation.

Therapeutic procedures in children with unilateral facial palsy are essential in minimizing consequences on occlusion and facial development. Facial palsy should be addressed as early as possible to restore optimal function as any disturbance of growth will result in alterations of jaw development.



Fig. 16. Shows three years post op. in animation.

Unilateral facial palsy during growth develops on asymmetric malocclusion with mandible deviation to the affected side because the unaffected side grows normally.

6*** Among the treatment options orthodontic therapy associated with physiotherapy has a good prognosis on symmetric development of the facial structures as described by this case report, this was not influenced by the nerve grafting as such

surgical procedure must be done by the end of the rapid development at the age of 13. The treatment should be addressed to promote the growth of soft and hard tissues.

Early diagnosis and functional treatment is essential as this can enhance opportunities for the clinicians to treat and solve any possible functional alteration, reducing invasive surgery to a minimum in order to improve the aesthetic needs.

The case report described in this paper demonstrates the positive prognosis that can be obtained with orthopedic treatment combined with physiotherapy exercises and reanimation surgery. In fact, the patient enjoyed complete recovery after treatment. The success was due to an early diagnosis and a III and IV degree of palsy. 3***.

A delay in the diagnosis could play a role in residual weakness of the face and mouth, but other factors such as severity of the inflammation and compression of the facial nerve are equally significant. Age and the degree of palsy are other reported prognostic factors; with younger patients and those with partial facial palsy the prognosis is better and they can reach full restoration [5,25,26].

4. Conclusions

Paediatric facial nerve palsy is a condition with severe implications due to the functional and aesthetic outcomes.

A careful differential diagnosis is recommended in order to establish the most appropriate treatment.

An early diagnosis and functional therapy is essential for reducing invasive surgery to a minimum, to improve symmetric development and the aesthetic needs which can't be restored surgically.

Hopefully regenerative medicine could offer in the future new options of the management of this condition.

Conflicts of interest

None.

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