External Mandibular Distraction in Unusual Pediatric Syndromes with Micrognathia and Airway Obstruction

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Received: 22 December 2010 / Accepted: 3 March 2011 / Published online: 25 March 2011
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Abstract Mandibular hypoplasia is a craniofacial malformation that can cause airway obstruction and may affect the eating process. This anomaly is common in many polimalformed Syndromes, including the Pierre Robin Sequence and the Moebius Syndrome. It is also common in cases related to the dysgenesis of the brainstem. In order to avoid tracheostomy, or to allow for early decannulation in severely affected children, external distraction devices were employed in 3 clinical cases to repair mandibular deficiencies. A successful external mandibular distraction induces the removal of the airway obstruction, allowing spontaneous ventilation without devices, an independent eating process with no need of nasogastric or gastrostomy tubes, apart from the linguistic and facial asymmetry improvement.

Keywords Mandible · Hypoplasia · Pediatric · Malformation · Distraction

Introduction

Several diseases affect craniofacial changes, such as the Pierre Robin Sequence, the Moebius Syndrome, Brainstem Dysgenesis, Collins Treacher Syndrome, Hemifacial Microsomia, Klippel-Feil Syndrome, Beck-Wiedemann

Syndrome, Trisomy 21, Freeman-Sheldon Syndrome, Craniocarpotarsal Dysplasia, Progressive Ossificans Fibrodysplasia, Vascular Malformations, Mucopolysaccharidosis, among others [1].

Mandibular hypoplasia is a craniofacial malformation which may cause airway obstruction and may cause difficulty in eating and consequently it can lead to death. Mandibular distraction represents a valid alternative to tracheostomy, as it prevents the airway obstruction, improving the breathing pattern as well as the patient's nutritional autonomy [2].

The sequence of Pierre Robin (SPR) is an autosomal recessive disease whose incidence is 1/5000–1/50000. This disease is characterized by the following triad: micrognathia (91.7% of the cases), glossoptosis (70–85%) and airway obstruction. Although the cleft palate is not included in this triad, it is commonly associated with this disease and it can worsen the obstruction. Furthermore we note that there may be difficulty in swallowing which causes a delay in growth and development. This disease has other features, including tracheomalacia, chronic aspiration, gastroesophageal reflux, defects in speech, obstructive sleep apnea and otitis media. Concerning systemic manifestations we may point out cardiovascular abnormalities, muscle-skeletal abnormalities, SNC abnormalities (delay in neurodevelopment, epilepsy, hypotonia) and genito-urinary defects. Most cases have a syndromic presentation (Sticker Syndrome, Treacher-Collins, Fetal-alcoholic). Only 17% are isolated (sequence of PR). The causes of SPR are intra-uterine pressure (caused by, for instance, oligohidramnios), family cases and failure in the intrinsic located mandibular growth [3–5].

The Moebius syndrome [6, 7] is a rare dominant autosomal disease, with an incidence of 2–20/1,000,000. It is characterized by congenital paralysis and non progressive ones of the VII and the VI cranial nerves (NC), associated with other abnormalities, especially in the limbs. It is noted

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a facial paralysis of the peripheral type, usually bilateral as well as convergent strabismus. When other cranial nerves are involved, unilateral or bilateral, we may detect a palpebral ptosis, divergent strabismus, deafness, disturbances of sensation in the territories innervated by the trigeminal nerve, dysphagia, dysphonia, tongue atrophy and difficulty in swallowing.

Abnormalities in the oral cavity may also occur: high palate, palatal cleft, micrognathia and hypoplasia of the enamel. Several patients show signs of mild mental retardation but in many cases speech problems and the absence of facial expression mislead the diagnosis. It was also noted cardiac malformations, central respiratory dysfunction, muscle-skeletal disorders (club-foot) and eye injury. The etymology, despite being speculative, is not only influenced by genetic factors but also by environmental ones, namely transitory fetal ischemic and hypoxic events, along with the use of drugs, such as misoprostol and thalidomide.

The designation Brainstem Dysgenesis is extended to patients with congenital dysfunction of cranial nerves and to the modification of the muscle tonus. In some cases, the dysgenesis is genetically determined and it may occur alone or be part of a polimalformative syndrome. Most cases are caused by lesions of vascular origin. Depending on the magnitude of the injury, the clinical symptoms can range from death in utero to small involvement of cranial nerves. The clinical findings in some of these patients may coincide with some described in the Moebius Syndrome, Pierre Robin or Cogan; it is important to consider a description of the clinical symptoms of the patient and/or of the segment of the brainstem affected.

Case Reports

The authors present 3 clinical cases of mandibular hypoplasia in the Pierre. Robin sequence, Moebius syndrome and Dysgenesis of the brainstem. In cases 1 and 3 children who had been submitted to a previous tracheostomy, showed no intubation problems, but in case 3, the child had ventilation issues. The child of case 2 was intubated without difficulty.

They all underwent a bilateral osteotomy at the angle of mandible and bilateral mandibular expansion with placement of small external nonresorbable mandibular distractors particularly designed for our department (Antão®). After a latency period of 3–5 days, mandibular distraction was activated and permitted unidirectional lengthening of 1 mm per day until maximal distraction was achieved (average: 19 mm). The distraction vector was oblique, and in a clockwise direction in relation to the occlusal plane. They were given 8 weeks for consolidation. Patients were subjected to a facial CAT (tomography) with 3D scanning in the pre and postoperative period (2 months long) in

order to confirm the wanted correction and to test the bone consolidation after the osteotomy.

Case 1

A three-year-old child, female, 18 kg, with a diagnosis of brainstem Dysgenesis, was admitted to the HSJ for mandibular distraction. This polimalformative syndrome affects the extra-pyramidal beam and the cranial nerves of 5–12 and it is associated with severe global delay of psycho-motor development and epilepsy, which is treated with sodium valproate.

The child has the following medical history: tracheostomy at 3 months old which resulted from respiratory failure, history of repeated respiratory infections in need of frequent aspiration of secretions; eating based on home cooking. After a physical examination the child showed signs of micrognathia and total hypotonia. 2 years ago she had initially undergone the setting of the resorbable internal device, which fell and cracked after a seizure. The patient was discharged after 4 days of hospitalization. Unidirectional mandibular lengthening was 17 mm, with functional improvement and a decreasing number of respiratory infections. The purpose to advance the mandible in this case was to increase the volume of the mandible and retroglossal oropharyngeal cross-section area, improving the upper airway volume and oxygen saturation, eliminating symptoms of obstructive sleep apnea. However, total hypotonia still remains a problem, and she needs a tracheostomy to maintain an open, functional airway.

Case 2

A 2-year-old child, female, with undefined weight and the bearer of Pierre Robin sequence had been submitted to a tracheostomy since birth because of respiratory failure refractory to glossopteris. She had a history of repeated respiratory infections and surgery to correct the palate cleft she had at 12 months of age. The physical examination revealed a prominent micrognathia causing further decline of the tongue with airway obstruction and difficulty in eating.

The child was discharged after 7 days of hospitalization. 6 months after the first surgical procedure (mandibular lengthening was 19 mm) a laryngeal—tracheoplasty was performed for the closure of the tracheostomy with costal cartilage grafting. Today the child shows a functional improvement in respiratory dynamics and language. About 2 months ago she had a spirometry, showing a normal respiratory pattern and values for her age (Figs. 1, 2, 3, 4).

Cinical Case 3

A 3-year-old child, male, 15 kg, is the bearer of chromosomopathy compatible with Moebius syndrome. Still in his

Fig. 1 Tracheostomized child with micrognathia—Pierre Robin Sequence

Fig. 2 Mandibular distractors

mother's womb he was diagnosed with polihidrâmnios. The child has a history of bicuspid aortic valve, obstructive sleep apnea (treated with noninvasive ventilation), kyphoscoliosis and an enlargement of the spinal canal. She had undergone a tonsillectomy with adenoidectomy in 2007 and a bilateral orchiopexies in 2008. The physical examination showed global hypotonia, bilateral facial paralysis, bilateral ptosis, emphasised retrognathism.

The surgery lasted 140 min of recording. In the post-operative process the patient was admitted to the UCI for 24 h and was extubated for 12 h after the surgery, having been discharged after 7 days of hospitalization. After a 9 month-follow-up we observe a significant improvement of the mandible, and no dependence on noninvasive ventilation. (Figs. 5, 6, 7)

Fig. 3 Results after 1 year

Sobrenome:		N.º Processo:	4014877
Nome:			
Data Nasc.:	27-04-2004	Idade:	4 Anos
Sexo:	feminino	Peso:	17,0 kg
Raça:	Caucasian	Altura:	103,0 cm
IMC:	16,02

		Prev	Pré	%Pré/Prev
FVC	[L]	0.93	1.05	113.0
FEV 1	[L]	0.82	0.88	108.1
FEV 1 % FVC	[%]	86.34	84.04	97.3
MMEF 75/25	[L/s]	1.21	0.89	73.6
MEF 25	[L/s]	0.74	0.50	66.7
MEF 50	[L/s]	1.46	0.99	67.7
MEF 75	[L/s]	2.09	1.29	61.8
PEF	[L/s]	2.20	1.33	60.5
IC	[L]	0.71		
IRV	[L]			
ERV	[L]	0.34		
VT	[L]	0.23		
MVV	[L/min]	15.81		
Date			22-12-08	
Time			13:48:18	

Fig. 4 Spirometry

Fig. 5 Micrognathia—Moebius Syndrome

Fig. 7 Results after 1 year

Fig. 6 Micrognathia—Moebius Syndrome

Discussion

The Pierre Robin sequence, Moebius syndrome and Dysgenesis of the brainstem have all in common the mandibular hypoplasia, which leads to a respiratory obstruction. The mandibular osteotomy and placement of nonresorbable distractors comes as a valid solution not only from the aesthetic point of view but also from the anatomical and functional point of view, as it brings an improvement of the airway, allowing, as it occurred in case 2, the closure of the tracheostomy, and in the case 3, avoiding the need to perform a tracheotomy.

First developed for orthopaedic surgery, distraction osteogenesis [8] has become an important way to repair severe mandibular deficiencies, being a safe, less-invasive procedure performed at any age for the repair of severe to moderate mandibular deficiencies.

Denny et al. [2] in 2001 demonstrated that mandibular advancement through distraction osteogenesis could

improve airway dimensions and resulted in decannulation in tracheostomy-dependent children.

Monasterio [9] and Sidman et al. [10], also showed excellent results using distraction to prevent the need for tracheostomy in neonates and children with mandibular hypoplasia with severe tongue-based obstruction. All of these authors used external distraction devices. These devices can be difficult to apply to the neonatal mandible and they inevitably result in scars as the distraction pins travel through the skin and it can become dislodged or infected during the course of distraction.

Bioresorbable internal distraction devices are not associated with the discomfort of wearing the external device, as there is less need for protection, no external skin scars. The most important advantage is that they do not require additional surgery for its removal, compared with internal and external metal devices [11, 12]. However, in our experience the complications included device failure to support resistance, and we can report a case of breakdown after an epileptic crisis (case 1). The main disadvantage is the size of current generation of bioresorbable material plate and screws which are larger in diameter and thicker in profile in comparison to titanium systems. Recently a new class of devices combined the features of an internal distraction device with resorbable plates and screws from LactoSorb (Walter Lorenz, Inc) including infant devices small enough for neonatal mandibular distraction.

We conclude that the cooperation of a multidisciplinary team is essential. This team should be made of Anaesthesiologists, Plastic surgeons, Otolaryngologists and Paediatricians to diagnose treat and prevent airway obstruction.

Conflict of interest None of the authors has a financial interest in any products, devices, or drugs mentioned in this manuscript.

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