

# Congenital oro-motor disorders

Children with congenital oro-motor disorders (COMD) are not numerous. However, as a group, they merit special consideration as management of diseases leading to COMD is complex, owing to their protracted and varying course, the diverse areas of health care involved, and the significant amount of resources their treatment requires.<sup>1</sup>

The term oro-motor disorders (OMD) refers to a group of diseases that predominantly affect sensory inputs, motor systems, and movement organization involved in sucking, chewing, swallowing, speech articulation, and non-verbal facial communication. Acquired diseases that selectively involve oro-motor functions in childhood are rare and, therefore, the number of patients reported has been scant.<sup>2</sup> Widespread congenital or acquired CNS lesions leading to severe forms of cerebral palsy, neurodegenerative diseases, or advanced neuromuscular disorders, in which oro-motor involvement is a relatively minor part of the patient's illness, are not generally considered under the heading of OMD.

Many aspects of COMD have been reviewed in the literature under different headings: congenital flaccid bulbar paresis,<sup>3</sup> dysphagia,<sup>4</sup> congenital suprabulbar and pseudo-bulbar paresis,<sup>5</sup> and feeding or swallowing disorders in children, which are the most commonly found titles in recent monographs or paediatric textbooks.

Preterm infants, and some term infants, may have delayed oro-motor function maturation and improve spontaneously.<sup>6</sup> Anatomical defects, either isolated or as part of polymalformative syndromes, are well-known causes of OMD; in some of the latter, however, it is likely that neurological factors also play a role in the oro-motor dysfunction. Widespread perinatal CNS insults can manifest as COMD in some patients. Corticospinal tract involvement largely improves during the recovery phase in these cases, leaving a residual and persisting suprabulbar or cerebellar dysfunction.<sup>4</sup> Congenital lesions with selective bilateral involvement of the perisylvian cortex or the cortico-bulbar tracts are associated with paresis of the tongue, soft palate, pharynx, and facial muscles. Patients with these anomalies, besides their OMD, are likely to present minor signs of pyramidal tract involvement, varying degrees of learning disability, and seizures.<sup>2</sup>

Recently, we proposed the term brainstem dysgenesis (BSD) to describe infants with congenital dysfunction of multiple cranial nerves and muscle tone due to prenatal lesions or anomalies of the brainstem.<sup>7,8</sup> Graham, in his paper on congenital flaccid bulbar palsy, anticipated this possibility,<sup>3</sup> and the series of Illingworth,<sup>4</sup> as well as Worster-Drought,<sup>5</sup> probably included children with BSD. In some patients, BSD is genetically determined and may be either isolated or part of a more extensive polymalformative syndrome.<sup>9</sup> In the majority of cases, however, prenatal destructive or disruptive lesions of vascular origin are the cause of this disorder.<sup>7,10</sup> The clinical manifestations in these patients, similar to what occurs with vascular accidents involving cerebral hemispheres, will depend

on the vascular territory affected and the extent of the brain tissue damaged.<sup>10</sup> In most patients described under Möbius, Pierre Robin, and Cogan syndromes, brainstem maldevelopment has been postulated<sup>7,8,11</sup> and, consequently, the nosology of these syndromes may be better approached if they are viewed as forms of BSD.

Finally, congenital myopathies, particularly those manifesting at birth and the congenital form of myotonic dystrophy, are the most common neuromuscular disorders that cause COMD. Initially, severe hypotonia, muscle weakness, and breathing difficulties play an important role in OMD. Later, velo-palatine insufficiency and anatomical changes in the oral cavity contribute to articulatory speech problems. Early recognition of these neuromuscular diseases is crucial, not only from the management point of view but also for family genetic counselling. Establishing the origin, nervous system location, and pathophysiology of diseases leading to COMD is an important issue since it provides clues to natural history and permits anticipation in terms of treatment and care provision.

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## References

- Clark M, Carr L, Reilly S, Neville B. (2000) Worster-Drought syndrome, a mild tetraplegic perisylvian cerebral palsy. Review of 47 cases. *Brain* **123**: 2160–2170.
- Christen HJ, Hanefeld F, Kruse E, Imhäuser S, Ernst JP, Finkenstaedt M. (2000) Foix-Chavany-Marie (anterior operculum) syndrome in childhood: a reappraisal of Worster-Drought syndrome. *Dev Med Child Neurol* **42**: 122–132.
- Graham PJ. (1964) Congenital flaccid bulbar palsy. *Br Med J* **2**: 26–28.
- Illingworth RS. (1969) Sucking and swallowing difficulties in infancy: diagnostic problem of dysphagia. *Arch Dis Child* **44**: 655–665.
- Worster-Drought C. (1974) Suprabulbar paresis. Congenital suprabulbar paresis and its differential diagnosis, with special reference to acquired suprabulbar paresis. *Dev Med Child Neurol* **16** (Suppl. 30): 1–33.
- Abadie V, Chéron G, Lyonnet S, Hubert P, Morisseau-Durand MP, Jan D, Manac'h Y, Couly G. (1996) Le dysfonctionnement néonatal isolé du tronc cérébral. *Arch Pédiatr* **3**: 130–136. (In French)
- Roig M, Gratacòs M, Vazquez E, Del Toro M, Foguet A, Ferrer I, Macaya A. (2003) Brainstem dysgenesis: report of five patients with congenital hypotonia, multiple cranial nerve involvement, and ocular motor apraxia. *Dev Med Child Neurol* **45**: 489–493.
- Roig M. (2005) Disgenesia troncocefálica: Los síndromes de Moebius, Cogan y Pierre Robin en revisión. *An Pediatr (Barc)* **62**: 347–351. (In Spanish)
- Holve S, Friedman B, Hoyme HE, Tarby TJ, Johnstone SJ, Erickson RP, Clericuzio CL, Cunniff C. (2003) Athabaskan brainstem dysgenesis syndrome. *Am J Med Genet* **120**: 169–173.
- Sarnat HB. (2004) Watershed infarcts in the fetal and neonatal brainstem. An aetiology of central hypoventilation, dysphagia, Möbius syndrome and micrognathia. *Eur J Pediatr Neurol* **8**: 71–87.
- Verzijl HT, Padberg GW, Zwarts MJ. (2005) The spectrum of Möbius syndrome: an electrophysiological study. *Brain* **128**: 1728–1736.