

Commentary: Pediatric ocular motor cranial nerve palsy: Demographics and etiological profile

The authors have presented an interesting article titled "Pediatric Ocular Motor Cranial Nerve Palsy: Demographics and Etiological Profile" on the etiology and clinical characteristics of pediatric III, IV, and VI cranial nerve palsies based on a retrospective study done in a tertiary care center in India. As mentioned by the authors, palsies of cranial nerves III, IV, and VI are rare in childhood, with an estimated incidence of 7.6 per 100 000 for all three palsies determined on a population-based study.^[1] There are limited studies on extraocular motor cranial nerve palsies in children in recent years. Also, most were

institution-based studies. Population-based studies derived from databases of epidemiological studies were rare.^[1]

The authors have showed that the most common extraocular motor cranial nerve palsy in their cohort was third nerve and the mean age of presentation was 7.02 ± 4.3 years. Homes *et al.* reported that fourth nerve palsy was the most common occurring in children.^[1]

Many large pediatric studies reported that the most common etiology of pediatric third (oculomotor) nerve palsy was congenital followed by trauma.^[1-3] The present study also reported similar findings. Neoplasm has been a rare etiology.^[1] Interestingly in a suprasellar teratoma third nerve palsy was noted after surgical resection. Aberrant regeneration which happens from misrouting of regenerating fibers is quite common

in congenital third nerve palsy, however primary aberrant regeneration seen in an acquired third nerve palsy in a child should raise concern for a compressive lesion with simultaneous ongoing repair. This has been reported in a cavernous sinus schwannoma or meningioma.^[4] Cyclical third nerve palsy has been reported in congenital third nerve palsy. Another etiology is third nerve dysinnervation that produces vertical and horizontal eye movement abnormalities together with ptosis and rare involvement of pupil. This is classified under congenital fibrosis of extraocular muscles (CFEOM1-3). The authors of the present manuscript also included other etiologies such as orbital apex soft tissue lesions, intracranial hemorrhage, and open lip schizencephaly. Despite pupil involvement, a compressive etiology is unlikely in children as compared with adults. Slow onset third nerve palsy and a progressive third nerve palsy in a child should prompt magnetic resonance imaging (MRI) of the brain with specific attention to the cavernous sinus.

Fourth (trochlear) nerve palsy is the commonest pediatric cranial nerve palsy.^[1] Due to a long intracranial course, this nerve is vulnerable to trauma. The most common etiology in children is congenital^[1,5] followed by trauma. Others are thought to be idiopathic. If congenital cases are excluded, trauma becomes the most common etiology.^[6] A bilateral case was seen in association with craniosynostosis. The present manuscript also found that isolated pediatric fourth nerve palsy was most commonly congenital in origin, acquired cases were usually idiopathic. Classical head postures include chin-down posture in bilateral cases and contralateral head tilt for unilateral cases. Usually, large vertical fusional amplitudes are seen in long-standing or congenital cases. Neuroimaging is generally low yield and is rarely needed. A high resolution 3 T MRI head may show absence of the fourth nerve in a congenital palsy suggesting superior oblique hypoplasia.^[7] Congenital fourth nerve palsy may be a form of congenital cranial dysinnervation disorder (CCDD).

Causes for sixth (abducens) nerve palsy in children include trauma, intracranial neoplasia (most often brainstem glioma), postviral, post-surgical resection of tumor, raised intracranial pressure, and congenital. Some have undetermined etiology. Important causes of congenital sixth nerve palsy include Duane's and Moebius syndrome, a form of CCDD. One series described trauma to be the most common etiology followed by neoplasia.^[5,6] The present paper also reported trauma as the most common cause followed by congenital, idiopathic, viral infection, presumed raised intracranial tension and neurosurgical complication. Holmes *et al.* felt majority of pediatric sixth nerve palsy were of undetermined etiology.^[1] Children with idiopathic sixth nerve palsy may be left with comitant esotropia. Neuroimaging has a high yield in pediatric sixth nerve palsy as it can recognize neoplasms and other important etiologies. Cases of sixth nerve palsy associated with neoplasia are generally not isolated.

The etiology of multiple cranial nerve palsies could be trauma, neoplasia, and aneurysm. Some studies report trauma to be the most common etiology,^[6] others report neoplasia as the common cause.^[5] This paper reports that combined cranial nerve palsy most commonly results from intracranial neoplasm followed by congenital, trauma and meningitis.

In summary, pediatric third, fourth, and sixth cranial nerve palsies are quite rare and are different from adults. The evaluation of pediatric extraocular motor nerve palsies also

should take a slightly different approach when compared to adults. It is important to recognize that the developing visual system in children with extraocular motor cranial nerve palsies is liable to develop strabismic, deprivation, or anisometropic amblyopia. The authors of this manuscript have also reported a high incidence of amblyopia that was noted in more than half of their patients. Amblyopia was most noted with third nerve palsy. There is loss of stereopsis and fusion but the ability to repair is also remarkably higher in children. Hence, it is important to intervene early and recognize and treat amblyopia. While not needed often, neuroimaging is likely to have high yield in specific cases of pediatric extraocular motor nerve palsies.

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