



Idiopathic Non-traumatic Facial Nerve Palsy (Bell's Palsy) in Neonates; An Atypical Age and Management Dilemma

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ABSTRACT

Idiopathic (Bell's) palsy is the commonest cause of unilateral facial paralysis in children. Although being idiopathic by definition, possible infectious, inflammatory, and ischemic triggers have been suggested. Bell's palsy is thought to be responsible for up to three-fourths of cases of acute unilateral facial paralysis worldwide. The diagnosis has to be reached after other causes of acute peripheral palsy have been excluded. However, it is rarely described in neonates and young infants. Steroids may have some role in treatment, but antiviral therapies have doubtful evidence of benefit. Prognosis is good, though residual dysfunction is occasionally encountered. We report the case of a two-week-old neonate with no prior illnesses who presented with acute left facial palsy. Clinical findings and normal brain imaging were consistent with the diagnosis of Bell's palsy. The patient had a good response to oral steroids.

Bell's palsy or, more precisely, idiopathic facial paralysis is one of the most common neurological disorders in children. The name came from Sir Charles Bell, who was the first author to describe unilateral facial weakness in 1830. The diagnosis can only be made after other causes of acute peripheral palsy have been excluded.¹ The role of steroids and antivirals in treatment of the condition is an ongoing debate.² Prognosis is good, though residual dysfunction may occasionally be seen.

As far as epidemiology is concerned, data varies between studies. It is estimated that the incidence of Bell's palsy is 15–25 per 100 000 people (including adults) per year.³ Bell's palsy constitutes 60–70% of patients with acute unilateral facial paralysis.⁴ Interestingly, it seems to affect the right side more frequently.⁴

Facial nerve paralysis in children might have variable etiologies apart from idiopathic Bell's palsy. Congenital, infectious, inflammatory, neoplastic, and traumatic causes are all important considerations before making a conclusive diagnosis of idiopathic Bell's palsy.⁵

CASE REPORT

A 15-day-old boy presented to our pediatric emergency center with a chief complaint of facial

asymmetry, which had lasted for two days. The mother noticed that he was unable to close his left eye. There was an obvious facial asymmetry, especially when he cried and some drooling of saliva from the left angle of the mouth. The family denied any history of fever, poor feeding, or hypoactivity. He remained a good sucker and active throughout his illness.

The mother had an uneventful pregnancy. She delivered at term via a normal, uncomplicated vaginal delivery. Her obstetric notes described the delivery as almost precipitate of short duration, and there were no clues of birth trauma. The infant's birth weight was approximately 3 kg, and he had normal initial Apgar scores after birth. There was no significant edema, caput, or cephalic-hematoma. His family was certain this facial asymmetry was not present in his first weeks of his life. There was no family history of note regarding neurological diseases.

Upon physical assessment, the patient was active and alert. He had a normal pulse rate and blood pressure readings compatible with his age. His head circumference was 34 cm with a normal head shape and flat anterior fontanel. He had positive red reflex, reactive pupils, and no obvious limitation in his eye movements. There was obvious facial asymmetry, with flattening of nasolabial fold in the left side, incomplete closure of his left eye, and deviation of the mouth towards the right side. All findings were

consistent with lower motor neuron left facial nerve palsy. His remaining neurological examination revealed normal spontaneous movement across all extremities with intact deep tendon reflexes. He had normally present symmetrical neonatal reflexes. No abnormal neurocutaneous stigmata were elicited.

His basic diagnostic workup was largely non-contributory. He had normal complete hemogram indices, serum electrolytes, urea, creatinine, C-reactive protein, glucose, ammonia, lactic acid, and blood gas figures. Cerebrospinal fluid (CSF) analysis was done. The results were negative for gram stain, bacterial growth in cultures, and viral polymerase chain reaction studies. He had normal levels of CSF sugar, protein, lactic acid, and cell counts. A consensus opinion was then to ask for brain imaging. Brain magnetic resonance imaging including special thin cuts in the temporal lobe, brainstem and facial nerve pathway was done and reported normal. The possible diagnosis of Bell's palsy was then entertained.

The patient was thus started on soluble oral prednisolone with a dose of 0.5 mg/kg/day for seven days. The patient had a remarkable rapid improvement and achieved a full recovery two weeks later. The baby was seen monthly over three months. He continued to have a normal neurological exam with full symmetry upon eye and mouth closure and opening, and when crying. With discussion and agreed assessment with physical therapist, it was obvious that there was no need for such support.

DISCUSSION

Bell's palsy is probably the most common mononeuritis in adults and children. Although it is an idiopathic disorder by nature, many possible triggers have been postulated. The exact pathophysiological pathways remain obscure and debatable. Some radiological findings have suggested that facial nerve compression, possibly secondary to ischemic injury, is the main pathological process as shown in facial nerve cuts demonstrating enhancement in some patients.⁶

Causes and triggers of facial nerve palsy have been elaborated extensively in the medical literature. The most popular theory was a link to the human herpesvirus (HHV). HHV was isolated in many patients with facial palsy in their endoneurial fluid after surgery.⁷ Other infectious agents have been assumed as well, including the influenza virus,

mycoplasma, syphilis, human immunodeficiency virus, and Lyme disease.^{8,9} A minority of children with Bell's palsy might have a positive family history of the same condition. Possible inheritance through the autosomal dominant route and incomplete penetrance has been suggested by some researchers.^{10,11}

Facial nerve palsy in young infants is rare and barely described in the medical literature. Neonatal facial palsy is almost always congenital and potential congenital malformation and traumatic causes have to be carefully sought. Traumatic etiology lies behind most cases of congenital facial palsy.¹² Risk factors for such trauma include pre- and post-maturity, being a first child, being overweight, and the use of forceps in delivery and delivery by cesarian section.^{13,14} The likely underlying pathological process is ischemic free radical-mediated tissue damage.¹⁵ Because of this uniquely small age group and the non-linear correlation with the injury type or duration, a full understanding of such pathophysiological mechanisms is unknown.¹⁶

Syndromic and genetic causes represent a small yet diverse group of etiologies associated with congenital facial palsy. It can be the result of central brain malformation syndromes, such as Arnold Chiari disorder, often along with other cranial nerves dysfunction.¹⁷ Facial palsy is one of the diagnostic criteria for Möbius syndrome, but it is usually bilateral.¹⁸ It has also been described in patients with branchial arch anomalies like Goldenhar, Poland, and branchiootorenal syndromes.^{19,20} Rarely, hereditary neuromuscular disorders can present with facial palsy that is noticeable at birth. Examples of such disorders include congenital myotonic dystrophy and congenital myasthenic syndromes.²¹ There is a current debate about new emerging genetic loci possibly responsible for a special disorder of hereditary congenital facial paralysis, which has been reported in a few families.²²

As far as idiopathic facial palsy in young infants is concerned, the literature is sparse. Manzouri et al,²³ described a three-month-old infant with acute unilateral facial nerve paralysis, assumed to have Bell's palsy after exclusion of other etiologies and who responded partially to oral steroids. McLellan et al,²⁴ were the first to report an acquired facial nerve paralysis in almost one-month-old infant in 1969, although no treatment has been offered and no data regarding the exact outcome of that case. In

the recent literature the only case of acquired facial nerve paralysis in a neonate was described by Saini et al,²⁵ in a four-week-old neonate with Bell's palsy and good clinical response to oral steroids. To the best of our knowledge our case is the youngest in history to be documented with acquired Bell's palsy.

Although facial nerve palsy is not the most common neurological disorder across the pediatric age group, the clinical burden of the disease is definitively heavy.²⁶ The significant impairment of functional and aesthetic aspects jeopardizes affected children's quality of life.²⁷ In spite of the relatively favorable natural history of this disorder, many caregivers do still consider it one of the true pediatric neurological emergencies particularly since it can interfere with breastfeeding; the primary activity at that age.²⁸

Clinical presentation is often obvious especially in older children with apparent asymmetry in the function of muscles responsible for facial expression. The peak of symptoms is usually reached 48 hours after onset. Physical examination is crucial to confirm the lower motor neuron nature of the facial nerve dysfunction and to try excluding other possible differential diagnoses. Signs become more evident when facial muscles are put into motor action, for example, when the child cries. There is no routine laboratory or imaging studies that are diagnostic, and the diagnosis remains solely clinical. House and Brackmann have a severity scale that is useful from a prognostic point of view and aids appropriate family counseling and further management.²⁹ However, this scale is difficult to implement in very young children.

The vast majority of children with Bell's palsy tend to recover without cosmetically obvious sequelae with or without medical treatment.³⁰ It seems that recovery three weeks from the onset is linked to more favorable prognosis helping the family counseling process.³¹ The old yet persistent debate about the role of steroids and antiviral therapy in children with Bell's palsy continues. The recent systemic review by Pitaro and his group of around 3 000 children with Bell's palsy concluded that evidence of the role of steroids is still inconclusive.³² The factor of time is crucial when there is a decision to treat though. There is a current cumulative evidence that steroids can be used if the patient presents in the first 72 hours of symptoms, and antiviral therapies should be used for patients with Bell's palsy grade four and above in the severity scale.³³ The debate extends to

other modalities of treatment like physiotherapy, laser therapy, and acupuncture as there is still no agreement about usefulness and safety of such interventions.³⁴ Obviously surgery is reserved for children with severe and persistent dysfunctional signs with variable outcomes.³⁵

CONCLUSION

Idiopathic Bell's palsy is a frequent clinical encounter in children and diagnosis has to be made with care excluding other causes of peripheral facial nerve dysfunction. Although there is no conclusive evidence about the usefulness of medical treatment, most children do achieve complete or near complete recovery. Bell's palsy is rare in young infants, and such a diagnosis should be entertained with caution in this young age group. We believe we are reporting the youngest neonate in the literature with idiopathic acquired Bell's palsy and good response to steroid therapy. This report highlights the importance of considering Bell's palsy as a potential diagnosis for facial nerve palsy in young infants, thus guiding further clinical care plans accordingly.

Disclosure

The authors declared no conflicts of interest.

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