Congenital Stridor Due to Bilateral Vocal Cord Palsy

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Abstract. Congenital stridor is one of the rare presentations of respiratory distress at birth. The commonest cause of congenital stridor is laryngomalacia, which accounts for 60% of the causes. The other common causes are congenital subglottic stenosis and vocal cord palsy (VCP). VCP is usually unilateral and most often linked with birth trauma, and is temporary. Bilateral palsy can be associated with other congenital anomalies. The current report describes a case of congenital bilateral VCP, not related to birth trauma and severe enough to require tracheostomy. [Indian J Pediatr 2005; 72 (5) : 443-444]
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CASE REPORT

A female baby was delivered vaginally to a primigravida at 37 weeks of gestation after an uncomplicated antenatal period. The baby developed stridor soon after birth which was present both during inspiration and expiration, high pitched and loud. Within four hours the baby developed hypoxemia (PaO₂ of 40mmHg) and hypercarbia (PaCO₂ of 70mmHg) on 100% oxygen. For this the baby was intubated and put on ventilation. The baby was weaned off ventilator within 24 hours. However, over next 24 hours she again developed CO₂ retention. She had good respiratory efforts. An endotracheal (ET) tube was inserted and left in situ. To exclude a transient palsy due to neuropraxia, an extubation trial was given on 16th day of life. However, this attempt failed and tracheostomy was done on 18th day of life. The baby also had hypoplasia of left lower limb below the knee. The measurements of right and left lower limbs revealed a difference of 2 cm in circumference and length. The baby developed myoclonic seizures on day eight, which responded to Phenobarbitone and clonazepam. Indirect laryngoscopy revealed minimal movements of both vocal cords suggestive of bilateral vocal cord palsy (VCP). To exclude surgical causes; investigations done included X ray neck, barium swallow, ECHO and MRI neck and head; all of which were essentially normal. Direct fluoroscopic laryngoscopy confirmed bilateral VCP. There was no instrumentation done during delivery and no other features of birth trauma were present. Metabolic work up including blood glucose, serum calcium, magnesium, blood gas, lactate and ammonia were essentially normal. Hence a diagnosis of idiopathic VCP was made. An EEG

done on Day 24 was grossly abnormal. Literature search did not reveal any syndromic relation between VCP, hypoplasia of lower limbs and seizures.

During follow-up the child was evaluated for neuromuscular disorders e.g. fasioscapulohumeral myopathy, spinal muscular dystrophy, and congenital myasthenia gravis. At one year of age, a trial of closing the tracheostomy tube for a few minutes was given but the child still needs the tracheostomy tube. There is no recurrence of seizures in the past 6 months and anticonvulsants have been stopped. Both mental and motor DQ's are mildly abnormal and have been assessed to be 70.

DISCUSSION

The most common causes of stridor at birth are laryngomalacia, congenital subglottic stenosis, VCP or vascular compression of trachea. The other causes are tracheomalacia, bronchomalacia and pressure effects of surrounding structures.¹ In a case series evaluating cause of stridor in babies it was found that Laryngomalacia (19.4%) was the commonest congenital cause.² In different case reports from India the other causes of stridor were tracheal duplication, ductus arteriosus aneurysm, subglottic cyst, tracheal and laryngeal agenesis and subglottic stenosis.³⁻⁶ Babies with stridor need to be referred to an otolaryngologist for full evaluation and treatment. The airway should be secured before referral.

Vocal Cord Paralysis (VCP)

It accounts for 10-15% of causes of stridor. Bilateral VCP can cause biphasic stridor. Unilateral VCP is more common and is usually associated with birth injury, aberrant vessels and atrial enlargement. Bilateral VCP causes more severe symptoms and aphonia.^{7,8} Often there

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is an associated neurological anomaly like Arnold chiari malformation, intraventricular hemorrhage, meningo-encephalocele and hydrocephalus. The other causes of VCP are traumatic forceps delivery, mediastinal surgery, and ligation of patent ductus arteriosus. However, most common cause of VCP is idiopathic.

Investigations which should be done to exclude other causes include a barium swallow, CT scan, MRI and endoscopy. Barium swallow helps to evaluate for aberrant vessels. CT Scan/ MRI head and neck can diagnose masses causing compression, aberrant vessels, mediastinal masses and associated neurological anomalies. Most infants with stridor require flexible direct laryngoscopy for diagnosis.⁹ Flexible fiber optic nasopharyngeal laryngoscopy is safe, allows detailed examination of nose, choanae, nasopharynx, oropharynx and hypopharynx.¹⁰ It provides information for evaluation of stridor, such as supraglottic soft tissues, vocal fold motion, dynamic compression from masses or cysts and evidence of trauma, tumor or infection. The dynamic view in particular is very helpful in diagnosing laryngomalacia and VCP. It can be done without anesthesia. Rigid endoscopic examination requires anesthesia.

Treatment of unilateral palsy depends on severity of symptoms. Most cases resolve over 6-12 months and hence surgical management is not required. Tracheostomy is often required in B/L VCP. Bilateral VCP related to birth injury may improve over two weeks in the presence of neuropraxia and 3-6 months in axonotmesis. Therefore, a trial of extubation at 14 days should be given in all cases of B/L VCP even if no obvious history of birth trauma is present. Additionally, in cases failing the extubation, a second trial should be given at 3-6 months. If the second trial also fails, then tracheostomy has to be continued and repair has to be attempted in later life.⁸ Rupa et al found that

tracheostomy was performed in 25% of cases with congenital stridor and the most frequent indication was bilateral abductor paralysis.² Another series showed that bilateral palsy had worst prognosis with only 52% recovering spontaneously and 7.2% requiring surgical correction¹¹

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