Etiology and management of bilateral vocal cord paralysis (VCP) in pediatrics differs from that in adults where it constitutes a part of the larger clinical entity called ‘bilateral vocal cord immobility’. Congenital laryngeal paralysis or bilateral abductor VCP presenting as neonatal stridor is rare and often requires prolonged mechanical ventilation. Syndromic as well as non-syndromic varieties of bilateral congenital laryngeal paralysis are documented in the Western literature. To the best of our knowledge, there are no reported cases from the Middle East. Here we report an Omani family wherein otherwise healthy and asymptomatic parents had three male newborn babies and one maternal uncle having features of congenital bilateral VCP suggesting X-linked recessive inheritance.

CASE REPORT
A full-term male newborn (weight 3.2 kg) was born by an elective cesarean section (indication: previous cesarean section) to a gravida-8, para-7 mother aged 32 years. The baby had severe inspiratory stridor and central cyanosis soon after birth. Endotracheal intubation was done given the worsening respiratory distress in the labor room and mechanical ventilation was subsequently initiated. Upon ear, nose, and throat evaluation with direct laryngoscopy, the vocal cords were found in the paramedian position with limited movements suggesting bilateral abductor vocal cord palsy. Flexible fiberoptic laryngoscopy was done to confirm the diagnosis. There was no evidence of laryngomalacia. The baby was ventilated for four days and weaned to nasal continuous positive airway pressure. Feeding was started on day two and full enteral feeds achieved by day six. Blood culture showed growth of Klebsiella pneumoniae and antibiotics were administered according to the sensitivity pattern (cefotaxime 150 mg/kg/day and amikacin 15 mg/kg/day). Additional investigations to evaluate congenital stridor including chest X-ray, cranial ultrasonography, echocardiography, skeletal survey, and routine laboratory work-up were normal. The baby had no dysmorphic features on physical examination; there was no evidence of any other organ dysfunction. Antenatal history for the present pregnancy was uneventful, and the mother had no existing or previously treated medical or surgical conditions.

Other causes of VCP like traumatic or forceps delivery, mediastinal surgery, and ligation of
patent ductus arteriosus, brainstem anomalies, and intracranial bleeding were not present in our case, and hence the diagnosis of idiopathic congenital bilateral vocal cord paresis was made. Significantly, there was a history of neonatal stridor in two previous siblings, both male. Both required respiratory support in the form of endotracheal intubation and mechanical ventilation for one and two weeks, respectively. The first child was diagnosed with bilateral VCP in India and no underlying defects were found on magnetic resonance imaging of the brain. Both had undergone fiberoptic laryngoscopy and respiratory symptoms resolved completely towards the end of first year of life without the need for tracheostomy as stated by parents. The elder siblings (aged six and 2.5 years, respectively) were asymptomatic with a normal voice. The parents had no respiratory symptoms, and there was no history of stridor at birth or dysphonia during infancy or childhood. A pedigree analysis for the family was done [Figure 1]. One of the maternal uncles of the index case had a history of noisy breathing at birth that resolved over several months according to the mother. We were unable to contact the uncle. The current child is being regularly followed-up. He is now eight months old and had normal vocal cord movements documented at six-months. His recovery was earlier than his elder siblings.

**DISCUSSION**

The diagnosis of idiopathic laryngeal paralysis in our case was straightforward as there was no history of a difficult delivery or other obvious causes for the presentation. There was a history of stridor at birth in two elder male siblings who were evaluated using laryngoscopy. The parents did not consent to any further investigations or invasive interventions (i.e., tracheostomy) in any child. However, the baby was successfully weaned from the ventilator as early as four days after birth. There were no congenital malformations or other comorbidities previously reported in the literature [Table 1].

VCP is the second most common cause (10–15%) of neonatal stridor, after laryngomalacia. However, there are fewer than 50 cases of neonatal presentations of bilateral ‘idiopathic’ congenital VCP identified in the English literature. Other causes of neonatal stridor are nerve injury during delivery, cardiothoracic surgery, and underlying neurological conditions. Radiographic evaluation including neuroimaging and flexible fiberoptic laryngoscopy should be done in all cases. Anomalies that need to be excluded in these cases include head and neck masses causing compression, Arnold Chiari malformation, meningoencephalocele, and hydrocephalus. Laryngeal electromyography wherein the electrodes are placed inside larynx is cumbersome in the pediatric age-group and therefore not tried in our case.

Infants with congenital VCP are usually symptomatic within the first 48 hours of life and often complicated by aspiration pneumonia. Tracheostomy is required in up to 70% cases with bilateral VCP but was avoided in our case. The

![Figure 1](image)

**Table 1:** Neonatal stridor due to congenital bilateral vocal cord paralysis: clinical associations reported in literature.6,7

<table>
<thead>
<tr>
<th>Isolated clinical comorbidities</th>
<th>Syndromic associations</th>
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<tr>
<td>Laryngomalacia</td>
<td>Robinow syndrome</td>
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<tr>
<td>Subglottic stenosis</td>
<td>Goldenhar syndrome</td>
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<tr>
<td>Subdural hematoma</td>
<td>Down syndrome</td>
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<tr>
<td>Central nervous system malformations including Arnold Chiari malformation</td>
<td>Williams syndrome</td>
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<tr>
<td>Arthrogryposis</td>
<td>DiGeorge syndrome</td>
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<tr>
<td>Facial dysmorphic features</td>
<td>Mobius syndrome</td>
</tr>
<tr>
<td>Veloopharyngeal insufficiency</td>
<td>Charcot-Marie-Tooth disease (type 1, 1b and 2)</td>
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<tr>
<td>Hypotonia/myopathy</td>
<td>JS-X syndrome</td>
</tr>
<tr>
<td>Ear deformities/sensorineural hearing loss</td>
<td>Hereditary neuralgic amyotrophy</td>
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<td></td>
<td>Congenital myasthenic syndrome</td>
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decision was based on parental concerns, as well as the fact that the baby could be weaned from ventilator early, and the siblings had a spontaneous recovery of vocal cord mobility. Several workers in the past have observed that only 35% of the children with bilateral VCP can be managed without tracheostomy.4,6,10 More recently, the trend is towards conservative management, especially in neonatal presentations.6,11 Recovery may take anywhere between six months and 11 years.10 Therefore, a waiting period of 12–18 months is recommended before contemplating definitive surgical intervention in children.

The earliest description of congenital bilateral vocal cord dysfunction by Plott dates back to 1964 and X-linked inheritance was postulated subsequently by other authors.12 There were no associated dysmorphic features in the original description. Association with psychomotor delay was probably due to severe hypoxic events during/after birth. Familial bilateral VCP in the absence of other associated features [Table 1] described in our report is not a well-known entity.5 It has aptly been called ‘Plott syndrome’ after Dwight Plott.7,13 Literature on clinical presentations and underlying genetic defects in congenital VCP are reviewed in a recent paper.14 The genetic basis for non-syndromic familial forms of VCP reported to date include a gene locus on 6q16 and inversion of chromosome 13 reported by Hsu et al.11b An extensive literature review on neonatal cases of congenital VCP is published.6 However, our literature review suggested that no cases of familial bilateral VCP have so far been reported from the Middle East, which makes our report unique. Four males in two successive generations in a family were affected at birth with a complete resolution of symptoms during infancy without complications like aspiration pneumonia, dysphonia, or long-term respiratory symptoms [Figure 1]. Tracheostomy seems to be unnecessary in familial non-syndromic cases (as observed in our case study) when compared to non-familial cases.11

We could not do genetic analysis in any of our patients due to cultural issues leading to reluctance to give consent. However, based on pedigree analysis, X-linked recessive inheritance is most likely, which is similar to the recent report of clustering in a family.5 Adult-onset bilateral abductor paralysis (Gerhardt syndrome) has also been reported in some families but is not X-linked.15 Moreover, the clinical presentation is at birth in Plott syndrome.

CONCLUSION

The diagnosis of bilateral VCP should be considered in cases with neonatal stridor. A detailed family history should be sought. Endoscopic laryngeal examination remains the diagnostic gold standard. In this report, we have described familial congenital bilateral vocal cord paresis with spontaneous recovery during infancy in an Omani family. The condition appears to be X-linked in inheritance.

Disclosure

The authors declared no conflicts of interest.

Acknowledgements

We thank the team of specialists in the ENT department for confirming our findings using fibreoptic laryngoscopy.

REFERENCES