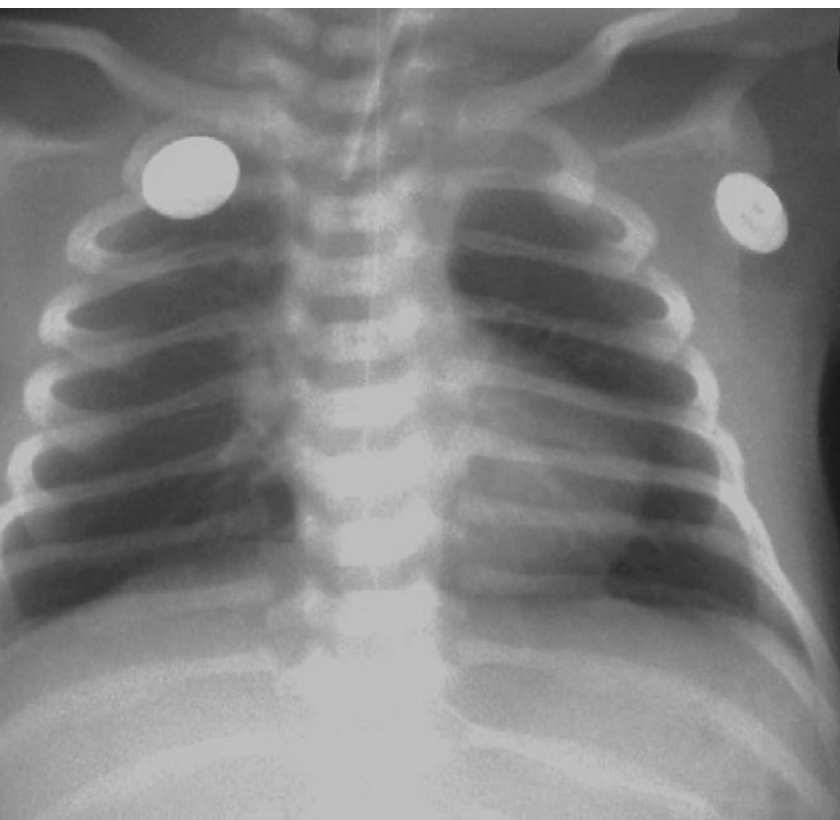


Congenital bilateral vocal
cord paralysis with unusual
clinical presentation

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Bilateral vocal cord paralysis in the neonatal period commonly presents with inspiratory stridor and respiratory distress of variable degrees. We describe an unusual case of vocal cord paralysis presenting immediately after birth with gasping respiration, bradycardia and negative pressure (i.e., *e vacuo*) pulmonary edema.

After uncomplicated pregnancy and labor, this male infant was delivered vaginally at term with an Apgar score 4, 5 and 6 at 1, 5 and 10 minutes, respectively. Umbilical cord pH values were 7.35 (venous) and 7.28 (arterial). Immediately after birth, severe gasping and bradycardia were observed. Cardiopulmonary resuscitation was initiated, followed by intubation which took place at the age of 50 minutes and was described by the anesthetist to be difficult. Thereafter the baby was transferred to our intensive care unit (ICU).

On admission to ICU (3 hours after birth), the baby was on conventional mechanical ventilation (CMV) (FiO_2 0.6, SaO_2 90-94%). Chest X-ray done soon after admission showed bilateral patchy attenuations (Fig. 1).

Echocardiography revealed tetralogy of Fallot with critical pulmonary stenosis, patent ductus arteriosus (PDA) with bidirectional shunt, and severe pulmonary hypertension. The patient was kept on CMV with

variable FiO_2 , and SaO_2 remained around 90%. Chest X-ray repeated at the age of 12 hours revealed dramatic clearance of lung fields and pulmonary oligemia (Fig. 2).

It was decided that the patient would need a Blalock-Taussig shunt, but that priority should be given to weaning from the respirator. Following extubation on day 8 of life, there was an increasing inspiratory stridor with moderate respiratory distress not responding to inhaled adrenaline. A formal evaluation of the airway was done by flexible endoscopy on day 15. It showed both vocal cords in a paramedian position without opening of the glottis during inspiration. The examination was done under inhalational anesthesia with sevoflurane. After endoscopy nasotracheal intubation was performed.

Due to decreasing oxygen saturations prostaglandin had to be started on day 17, and on day 18, a modified 4 mm Blalock-Taussig shunt was created. Finally, after complete wound healing, tracheostomy was done on day 29.

Family history in the present case is of particular interest. The family is originating from Turkey, and there is consanguinity of the parents (Fig. 3). In addition, an uncle was treated in our hospital 25 years ago for congenital bilateral vocal cord paralysis (tracheostomy up to the age of 6 years). There are also many unexplained neonatal deaths in the family.

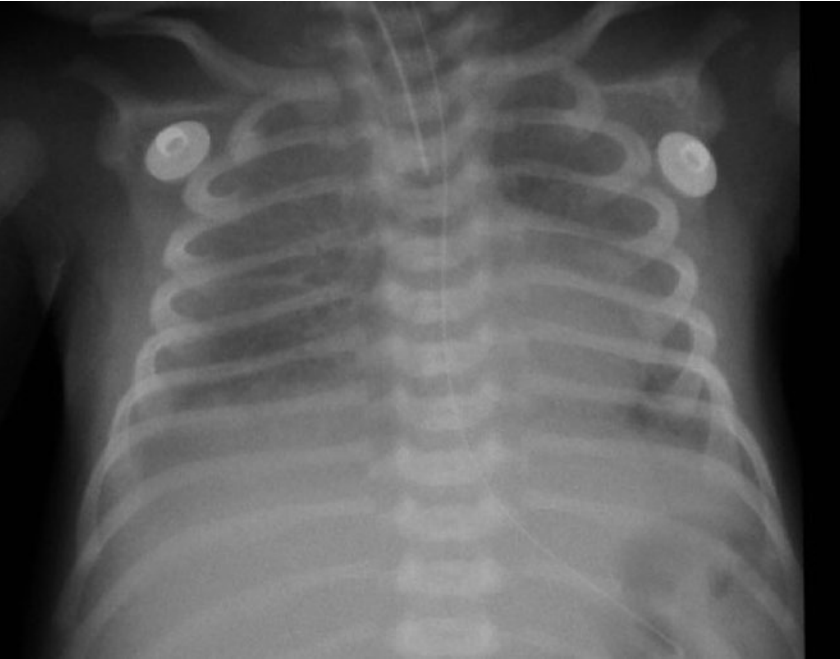


Fig. 1

Chest radiography 3 hours after birth showing bilateral patchy attenuation (pulmonary edema e vacuo).

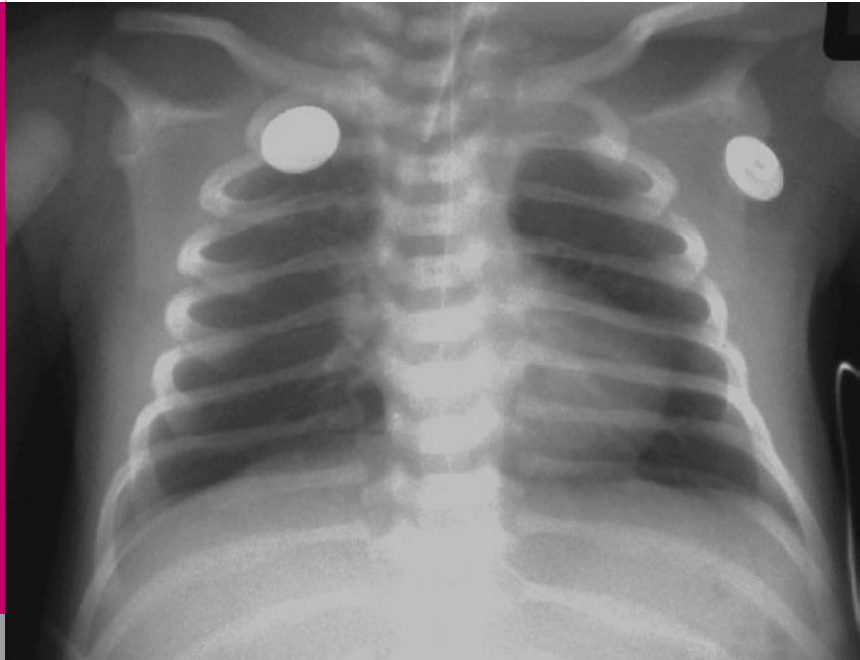
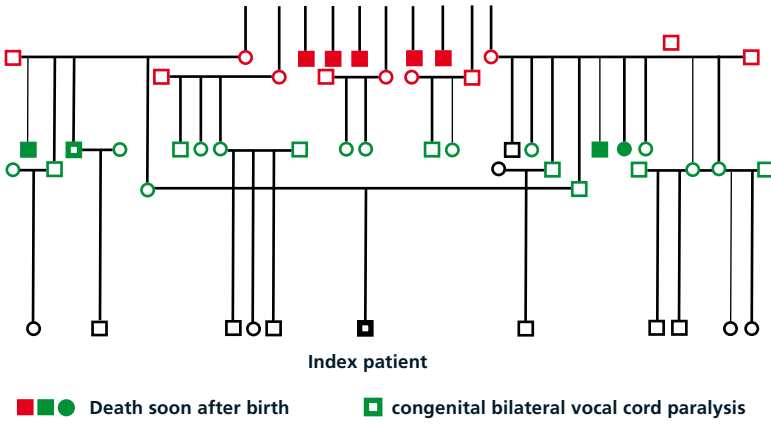


Fig. 2

Chest radiography 12 hours after birth with complete clearing of pulmonary edema and signs of pulmonary oligemia.



Pedigree of the patient's family.

Fig. 3

DISCUSSION

Vocal cord paralysis accounts for 10% of all congenital laryngeal lesions and is, after laryngomalacia, the second most frequent cause of neonatal stridor. An overview of etiologies of vocal cord paralysis in all pediatric age groups is given in the table.

CNS: central nervous system (Arnold-Chiari malformation, leukodystrophy, encephalocele, myelomeningocele, hydrocephalus, cerebral or nuclear dysgenesis); PNS: peripheral nervous system; ETT: endotracheal tube; VCP: vocal cord paralysis; VSD: ventricular septal defect.

Table

Etiology	Unilateral VCP	Bilateral VCP
CNS (most brainstem)	Infrequent	Common
PNS	Infrequent	Common (myasthenia gravis)
Trauma	Common (thoracic surgery, ETT)	Infrequent
Neoplasm	Common (skull base tumor)	Infrequent
Inflammatory	Infrequent	Infrequent
Cardiovascular anomaly	Common (VSD)	Infrequent
Metabolic	Infrequent (chemotherapy)	Infrequent
Genetic	Infrequent	Infrequent
Idiopathic	Common	Common

Familial vocal cord dysfunctions are rare (2-4) and most often isolated. The mode of inheritance is variable and has been reported to be X-linked, autosomal dominant or recessive. With regard to the present case we assume a genetic cause, most likely with X-linked inheritance. Chromosomal analysis and neuroimaging were both normal.

Bilateral vocal cord paralysis commonly presents with inspiratory stridor, unilateral paralysis with hoarseness. The unusual clinical presentation in our case with severe respiratory distress was most likely due to pulmonary edema. Rapid clearing of the lung fields after relief of upper airway obstruction and positive pressure ventilation makes this hypothesis very plausible. The fact that this occurred in a neonate with severe tetralogy of Fallot (severe pulmonary hypoperfusion) is astonishing, however, the particular situation of transitional circulation might have produced enough pulmonary blood flow through the patent ductus arteriosus to allow for development of pulmonary edema. Negative pressure pulmonary edema is well known in the pediatric literature (5-7). It is thought to be due to high pressure gradients between pulmonary capillaries and airspaces when the patient inspires against an obstructed upper airway.

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