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Case report

Isolated idiopathic bilateral vocal cord paralysis in two sisters: Case report and review of familial vocal cord paralysis

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ABSTRACT

Two sisters noted to have neonatal stridor were diagnosed with isolated idiopathic bilateral vocal cord paralysis. The vocal cord paralysis spontaneously resolved in both after being tracheostomy tube-dependent for varying periods of time. This is the third known isolated case documented in sisters. An updated literature review of this uncommon complex condition is presented.

1. Introduction

The second most common cause of stridor in infants is vocal cord paralysis (VCP) [1]. VCP in the pediatric population can be classified as unilateral or bilateral. Unilateral VCP is more common, associated with iatrogenic causes, and patients being asymptomatic. Bilateral VCP (BVCP) is most often idiopathic and associated with more pronounced obstructive airway symptoms. A high proportion of patients with BVCP will require a tracheostomy [2].

VCP is being diagnosed more frequently due to increased recognition, advances in diagnostic techniques, and improved survival of premature infants [3]. Familial cases of VCP have been described infrequently. Their occurrence indicates a genetic component to normal laryngeal development and function [2].

This paper will present a case of two sisters noted to have stridor at birth who were eventually diagnosed with isolated idiopathic BVCP requiring tracheostomies. An updated review of the current literature on familial vocal cord paralysis will also be provided (Table 1).

2. Case studies

2.1. Case 1: A.T.

A.T. was born to non-consanguineous parents at term by an atraumatic vaginal delivery. At birth she was noted to have inspiratory stridor. She was subsequently seen in the ENT clinic at 6 days of life (DOL). She was initially diagnosed with laryngomalacia as the cords could not be visualized on flexible fiberoptic laryngoscopy (FFL). A rigid bronchoscopy and CO_2 laser supraglottoplasty were performed on DOL 26. Her symptoms failed to improve after the procedure and on repeat FFL she was found to have BVCP. Magnetic resonance imaging (MRI) of the head, neck, and upper thorax were normal. Direct laryngoscopy and tracheostomy were performed at 6 weeks of age. At 4 months of age routine FFL examination demonstrated a return of function in her vocal cords. She was successfully decannulated at 5 months of age.

2.2. Case 2: N.T.

N.T. is the younger sister of A.T. by 2 years. She was the product of an uncomplicated pregnancy and vaginal delivery. She was noted to be stridorous at birth, requiring bag and mask ventilator support. She was transferred to the Neonatal Intensive Care Unit (NICU) for ENT evaluation. She was found to have laryngomalacia and normal vocal cord mobility. At 2 months of age repeat FFL revealed BVCP. MRI of the head, and computed tomography (CT) of the chest and neck failed to reveal any abnormalities. She was admitted to hospital for rigid bronchoscopy and tracheostomy and

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Table 1
Case reports of familial vocal cord paralysis.

Author	Affected family members	Associated abnormalities	Most likely mode of inheritance	Spontaneous recovery
Isolated Idiopathic BVCP				
Mace et al. [8]	4 m, 1 f – over 3 gen	None	AD	N
Brunner and Herrmann [9] (*German paper*)	Mother, son, 2 dghtrs	None	AD	?
Cunningham et al. [10]	Brother, 2 sisters	None	AD	Y
Isaacson and Moya [11]	Mother, dghtr	None	AD with new mutation	Y
Manaligod and Smith [2]	M infant, 2 f maternal cousins	None	AD with variable expressivity	Y
Raza et al. [12]	Brother, sister	None	AR – 1st degree parental consanguinity, paracentric balanced inversion of ch13 in mother and 2 sibs	Y
Khodaei et al. [13]	2 brothers	Younger bro had LTM, marked reflux laryngitis, epilepsy	? Difficult to ascertain	Y
Tarin et al. [14]	Father, son	None	AD with variable penetrance	Y
Omland and Brondbo [15]	Twin boy and girl	None	AD	N
Idiopathic VCP associated with anomalies or an underlying genetic syndrome				
Plott [5]	4 brothers	MR, generalized neuromuscular incoordination, inner ear deafness in 1	XR	Y 1 death
Watters and Fitch [16]	2 brothers, 1st cousin once removed	Severe MR, psychomotor retardation, blindness in 1 bro	XR	? 1 death
Gacek [17]	Father, 2 sons	Motor nerve deficits of UE and LE	AD	N
Hollinger et al. [18]	Mother, son	CMT	?	N
Young and Harper [19]	9 family members – 3 m, 6 f	Insidious onset of progressive, chronic distal spinal muscular atrophy and weakness	AD	N
Morelli et al. [20]	7 (4 m and 3 f) across 3 generations	2 with clubfoot	AD	N
Grundfast and Milmoe [21]	Father, son, dghtr	Swallowing difficulty during infancy	AD	Y
Tucker [22]	Father, dghtr	Bilateral ptosis of eyelids at birth Prematurity, ?mild MR in dghtr	Possibility of new sx complex of congenital origin	N
Serratrice et al. [23] (*French paper*)	Brother, sister	Proximal and distal atrophy and proximal muscle weakness consistent with chronic spinal muscular atrophy	AD	?
Boltshauser et al. [24]	Mother, dghtr, grandfather	Progressive distal spinal muscular atrophy and weakness and SNHL	AD – pleiotropic effect of a single mutant gene	N
Hawkins et al. [25]	Mother, identical m twins, m sibling, 1st m cousin	Digital anomalies	AD with variable penetrance and male predilection – single inherited trait	Y
Schinzel et al. [26]	Mother, son, 2 dghtr	Moderate MR, youngest girl was microcephalic	AD with variable expression – remotely consanguineous, parents	Y for 2; N for 1
Pridmore et al. [27]	7 family members in 3 successive gen (3 m and 4 f)	Progressive distal spinal muscular atrophy and weakness	AD	N
Dyck et al. [28]	2 kindreds with 14 pt 8/11 in kindred 1 had sx of VC/resp involvement. (6f, 2 m). Kindred 2 had 2/3 (2f) affected	Variable degree of muscle weakness of limbs, vocal folds, intercostal muscles, a sx sensory loss CMT II-C 5 had VCP	AD	N
Koppel et al. [7]	2 brothers	Swallowing problems, delayed neurodevelopment	AR – parents are double 1st cousins, maternal grandparents are first cousins and paternal grandparents are 2nd or 3rd cousins	N
Lacy et al. [29]	Mother, son, dghtr	CMT II-C Son had normal intelligence, deep inspiratory stridor, moderate pectus excavatum, peroneal atrophy, weakness in all 4 extremities. Dghtr had weakness of instrinsic muscles of hands and bilateral dorsiflexion of feet and toes	AD	N
Cuesta et al. [30]	2 small families and 1 large inbred Spanish family	Axonal CMT Childhood onset Weakness, foot and hand wasting	AR	N