Double Aortic Arch in d-Transposition of the Great Arteries Complicated by Tracheobronchomalacia

Stanislav Tůma,1 Zdeněk Slavík,2 Petr Tax,2 Bohumil Hučin,2 Jan Škovránek2
1 Clinic for Imaging Methods, University Hospital Motol, CZ-150 18 Prague, Czech Republic
2 Center of Pediatric Cardiology and Cardiac Surgery, University Hospital Motol, CZ-150 18 Prague, Czech Republic

Abstract
Simultaneous occurrence of d-transposition of the great arteries and aortic arch malformations is very rare. A case of this malformation, the fourth case reported in the literature, is described. Despite successful surgery, tracheobronchomalacia was fatal at the age of 7 months.

Key words: Double aortic arch—d-transposition of the great arteries—Tracheobronchomalacia

The simultaneous occurrence of d-transposition of the great arteries and aortic arch malformations is very rare. We report a case of double aortic arch associated with d-transposition of the great arteries which, to the best of our knowledge, is the fourth case reported in the literature [1, 2].

Case Report
The patient, a male, was born May 7, 1986 after a complicated delivery at term. Severe cyanosis was evident from birth. A systolic regurgitant murmur and right ventricular hypertrophy were additional findings present on admission to a specialized supraregional medical center the same day.

An echocardiogram revealed visceral situs solitus with atroventricular concordance and ventriculoarterial discordance, supporting the diagnosis of transposition of the great arteries. A small ventricular septal defect, patent foramen ovale, and patent ductus arteriosus were also present.

Balloon atrial septostomy was performed 24 hours after birth; angiocardiography at the same time confirmed the previous diagnosis but also showed a double aortic arch (Fig. 1). Repeat echocardiogram and esophagogram (Fig. 2) clearly demonstrated the anomaly of the aorta.

Mild mixed stridor, present on admission, quickly progressed and resection of the left-sided aortic arch, distal to the origin of the left subclavian artery, along with resection of bilateral ducts were required at the age of 3 weeks. Ventilation was difficult and tracheobronchomalacia was fatal at the age of 7 months. Tracheobronchography (Fig. 3) after the second operation showed severe bilateral tracheobronchomalacia. The infant could not be weaned from the ventilator and died 6 months later of severe pneumonia due to pseudomonas infection.

Discussion
Transposition of the great arteries is characterized by discordant arterial connection. The origins of the aorta and pulmonary artery are reversed: the aorta arises from the right ventricle ("d" refers to dextroposition) and the pulmonary artery arises from the left ventricle. Anomalies of the aortic arch are very rare under these morphologic and hemodynamic conditions. Aortic arch anomaly or variation was found in 16 patients (28.1%) in a series of consecutive patients investigated for transposition of the great arteries [3]. The combination of d-transposition with double aortic arch is extremely rare. Higashino and Ruttenberg [1] reported a case in which the diagnosis was made only by necropsy and had not been seen in a previous angiocardiogram. Kupferschmid et al. [2] reported a successfully diagnosed malformation in a 1-day-old girl, made by angiocardiography at the time of balloon atrioseptostomy which was confirmed later by echocardiography. The child was alive after surgery for interchange of the great arteries, closure of atrial and ventricular septal defects, dissection of the left patent ductus arteriosus, and division of the right aortic arch distal to the origin of the right subclavian artery [2]. She was, however, the only surviving patient. Both papers [1, 2] cited the first case described in 1837 by von Siebold. Our report is the fourth case described in the literature (Table 1) [1, 2].
Pressure-induced ischemia of the tracheobronchial wall, stepped up by hypoxemia and unusual position of the great vessels, probably contributed to the severe tracheobronchial damage in our patient. The extent and degree of tracheal and/or bronchial damage clearly limits the success of the treatment. Recent results in surgical tracheal reconstruction in a selected group of patients with tracheomalacia are encouraging. Conservative treatment quite frequently requires tracheostomy and long-term ventilation. The evidence for successful growth of airways with malacia in infancy remains anecdotal. Expandable metallic stents were used in experimental and clinical applications at that time, but were not common. Even now, stenting of short segments of airways has shown some optimistic midterm results [4], but long-term experience and well-tolerated, expandable and/or removable stents are still needed, especially for growing structures of children. Aortopexy is a successful method for treating tracheomalacia due to the anterior crossing of the trachea by the brachiocephalic trunk [5] and was not indicated in our case.

The severity of tracheobronchial damage in our patient did not allow stenting or surgical reconstruction.