period as they were considered to have a heart malformation because of the unusual electrocardiographic finding. It is speculated that these forms of congenital left axis deviation, in absence of other cardiac malformations, could form a part of those left axis deviations which, till now, when observed in adults, have perhaps been erroneously labelled as "acquired" conditions.

Three of our cases had a history of a spontaneously closed ventricular septal defect which could form one possible explanation for this congenital idiopathic left axis deviation. Only two of them also had an incomplete right bundle branch block.

A striking feature from our data is the apparently complete sex dependence of this electrocardiographic condition in childhood. This observation is in disagreement with the findings of Gup et al. [4]. In their paper, examining a population that was 1/4 of ours and without infants, the sex was equally distributed. No information about male prevalence has been given in other clinical reviews.

In conclusion, congenital forms of left axis deviation can exist in otherwise normal hearts. They can probably be masked in the early days of life, while the right ventricular predominance balances the initial left ventricular forces. Recognition in the newborn and childhood period is very possible with routine electrocardiographic examination. The association with a spontaneously closed ventricular septal defect, as well as our observed male prevalence, need further confirmation and evaluation.

References


Echocardiographic diagnosis of interrupted aortic arch with an aortopulmonary communication

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The association of interrupted aortic arch and an aortopulmonary communication is extremely rare and represents a diagnostic challenge. We report the case of a 15-hour-old infant presenting with these malformations with emphasis on the echocardiographic diagnosis. We conclude that cardiac catheterisation is not mandatory and indeed, is best avoided.

Key words: Interrupted aortic arch; Aortopulmonary communication; Echocardiography

Introduction

Interrupted aortic arch is a rare congenital malformation which, in extremely rare instances, may be asso-
associated with an aortopulmonary communication [1,2]. The association of these two malformations has been recently reviewed [2], and the majority of cases were the results of autopsy investigation. In contrast, our knowledge of echocardiographic diagnosis of this condition is limited [3]. In this report we describe a neonate who underwent corrective surgery without the need for invasive investigation.

Case Report

A full-term male infant was delivered with an Apgar score of 9 after 1 minute. His birth weight was 3.4 kg. Tachypnoea and cyanosis were noticed six hours after delivery. At 15 hours of age, the femoral pulses disappeared and coarctation of the aorta was suspected. The infant rapidly developed severe heart failure with pulmonary oedema. The brachial pulses were normal and symmetrical, with an upper limb blood pressure of 80/40 mm Hg. The apical impulse of the heart was hyperkinetic, the first heart sound was normal and the pulmonary component of the second heart sound was loud. No click nor murmur was heard. The liver was palpable 4 cm below the costal margin. The biochemical and haematological laboratory results were normal. The arterial blood gases (taken from umbilical artery) on FiO₂ 50% showed a pH 7.25, PCO₂ 45 mm Hg, PO₂ 53 mm Hg, and O₂ saturation 82%. The chest roentgenogram showed cardiomegaly and pulmonary oedema. The electrocardiogram was normal for age. Cross-sectional echocardiography was performed using a Diasonic CV 400 with a 5 MHZ transducer. The intracardiac anatomy was normal. There was no ventricular septal defect. The size of the ventriculo-aortic junction and the proximal ascending aorta were normal. The pulmonary valve and its origin from the right ventricle were normal in size and location. The take-off of the coronary arteries was normal. At about 5 mm above the orifice of the left coronary artery, a large (15 mm) aortopulmonary communication was seen running posteriorly from the left and posterior aspect of the ascending aorta up to the bifurcation of the pulmonary trunk (Figs. 1, 2). The communication was asymmetrical (6 mm anteriorly and 3 mm posteriorly). Its pathway was visualised in all standard echocardiographic views although the proximal portion was best demonstrated using the apical 4-chamber view while the distal connection to the pulmonary arteries was best seen using the subcostal long and short axis views as well as the parasagittal views. The segment of the ascending aorta distal to the aortopulmonary communication was narrow (4–5 mm). The arch was interrupted just distal to the left subclavian artery as revealed by suprasternal and parasagittal views (Fig. 2). The descending aorta was connected to the main pulmonary artery by a large (8 mm) arterial duct which permitted a predominant

![Echocardiographic apical 4-chamber view and diagram showing the take-off of the aortopulmonary communication from ascending aorta.](image)
right-to-left shunt as detected by Doppler echocardiography.

Following the echocardiographic diagnosis, the infant underwent surgical repair which included direct anastomosis of the arch, ligation of the aortopulmonary communication and division of the arterial duct. A residual 40 mm Hg gradient from the arch to descending aorta was recorded following the anastomosis so a 6 mm “Gore-tex” conduit was interposed from the left common carotid artery to the descending aorta. One year after surgery there is no haemodynamic abnormality.

Discussion

Interruption of the aortic arch with an aortopulmonary communication is rarely reported and is observed in about 0.3% of congenital heart malformations. As emphasized by Braulin et al. [2], who reviewed 46 cases (31 from the literature and 15 collected from multiple centres), the delineation of anatomical details in this condition is important since marked variations in anatomy are not uncommon [4,5].

The purpose of this communication is not to add a new case report to this rare entity, but mainly to demonstrate the ability of cross-sectional echocardiography to diagnose correctly the detailed anatomy. Moreover, in such a condition in which the left ventricular function is jeopardized, prominent pulmonary oedema and the impaired renal function render cardiac catheterisation extremely hazardous and favour a non-invasive diagnostic investigation.

The intracardiac anatomy, the pathway of the communication and the arch anatomy were readily visualised using standard and modified echocardiographic views. The absence of a ventricular septal defect and confirmation of the normal distribution of the coronary arteries, emphasised in the “rule of thumb” for preoperative evaluation [2], were also well demonstrated. Angiographic confirmation is not needed and, indeed, should be avoided.

References

Tricuspid and pulmonary atresia with coarctation of the aorta: a rare combination possibly explained by persistence of the fifth aortic arch with a systemic-to-pulmonary arterial connection

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Introduction

The rare association of tricuspid and pulmonary atresia with hypoplasia of the transverse aortic arch complicated by the unique finding of a persistent fifth aortic arch with a systemic-to-pulmonary artery connection on the same side as the definitive aorta. The histology of the fifth aortic arch is consistent with the pattern of an arterial duct.

Key words: Coarctation of the aorta; Congenital heart disease; Ductus arteriosus; Embryology; Fifth aortic arch; Tricuspid atresia

Case Report

A ten-day-old baby boy with a birthweight of 3700 grams was admitted to the Niigata University Hospital with severe cyanosis, dyspnea, and peripheral edema. The heart rate was 160 and the respiratory rate 80/minute. The blood pressure in right arm was 85/50 and the femoral pulses were palpable. The precordium was quiet. The second heart sound was single; and a grade 1/6 systolic murmur was noted along the left sternal edge. The liver was moderately enlarged. The chest radiograph demonstrated minimal cardiac enlargement and oligemic lung fields. An echocardiogram

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