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CASE HISTORY

Congenitally corrected transposition of the great arteries

Jacqueline Smith*, **Ann Sproul**, **David Watson**, **Yoga Kandasamy**

The Townsville Hospital, Neonatal Unit, Angus Smith Drive, Douglas, Townsville, Queensland, Australia

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Case report

Our female baby S was born at 39 + 1 weeks by spontaneous vaginal birth (SVB) to a 21 year old Papua New Guinean mother, in her first pregnancy. Baby developed cardio- respiratory compromise at birth and required intermittent positive pressure ventilation (IPPV) via bag and mask for two (2) minutes before her heart rate increased to >100 beats per minute. Her color, which was initially dusky, pinked up and her perfusion improved prior to her transfer to our special care nursery for ongoing observation for an antenatally diagnosed cardiac anomaly. Her birth weight (BW) was 2.97 kg, head circumference (HC), 34 cm and length was 47.5 cm; all being appropriate for gestational age. Maternal serology was unremarkable, with an O positive blood group. However, her Group B Streptococcus (GBS) status was unknown. Her mother had developed pyrexia during labor, which could have been attributed to a prolonged second stage of labor or to that of a septic nature (Chen et al., 2012). A septic screen was performed and penicillin and gentamicin with antifungals were given until blood cultures were available at 48 h of incubation. These remained negative and the antibiotics were ceased.

Antenatal ultrasound at 36 weeks identified ccTGA (see Fig. 1). The cardiac apex was positioned to the left, with situs solitus, however there was atrioventricular discordance and a large VSD. The aorta arose from the morphological right ventricle (on the left). The pulmonary artery arose over the ventricular septal defect. The morphological left ventricle was right sided. The left sided AV valve (tricuspid) straddles the large VSD. The outflow tracts did not appear obstructed. These findings were consistent with anatomically corrected transposition of great arteries and a large VSD. A repeat echocardiogram at day four of life replicated these findings. During her stay in the unit she was observed for any signs of cardiac failure with saturation and cardiac monitoring and for visual signs of respiratory distress. Baby S was commenced on enteral feeds from day one of life and established full breastfeeds within the first few days. She was discharged home close to two (2) weeks of age, thriving well with a weight increase of 200 g; testimony to her good progress. Her follow up would entail visits to one of our

* Corresponding author.
E-mail address: jackiesmith3@me.com (J. Smith).

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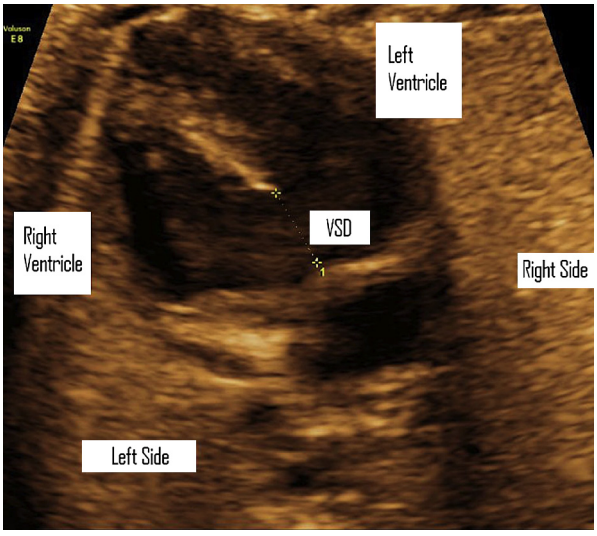

Fig. 1 Fetal ultrasound showing ccTGA. Image taken by Dr David Watson, Fetal and Maternal Medicine, The Townsville Hospital, Queensland, Australia.

neonatologists for growth and development reviews and visits to a consultant cardiologist.

A copy of her birth details and a summary of her stay in the unit were also sent to the general practitioner and child health nurses. This ensured the parents had a multidisciplinary support network, both at the hospital and in the community, should any problems occur. The parents were also provided with an information sheet on signs of heart failure and were advised to bring their daughter back into the hospital for review if she developed any of these signs or they were concerned about her.

Introduction

Congenitally corrected transposition of the great vessels (ccTGA) is a rare congenital heart defect, seen in $<1\%$ of those with congenital heart disease (See Fig. 2). It is characterized by atrio-ventricular (AV) and ventriculo-arterial (VA) discordance, whereby the receiving chambers (atria) are connected to their opposite pumping chambers (ventricles), due to a swap over from their normal anatomical positions during embryonic development (Rummell, 2011; Flack and Graham, 2012). This means that the normally (see Fig. 3) placed aorta arises from the inverted right ventricle, which is now positioned on the left and that the normally placed pulmonary artery stems from the inverted left ventricle, now taking its position in the right side of the heart. It carries de-oxygenated blood to the lungs. The consequences of this inversion sees the de-oxygenated blood on the right side of the heart being channeled to the lungs for oxygenation and returned to the left side via the pulmonary veins, to be pumped systemically via the tricuspid valve. This valve, which is normally sited in the right side of the heart, is designed for low pressure pumping but is now under great stress as it sends oxygenated blood to all areas of the body, making this valve extremely susceptible to regurgitation (Warnes, 2006). The usual pathway of blood flow continues as normal. Hence, this condition's nomenclature being that of 'corrected'.

In contrast to this anomaly is transposition of the great arteries (TGA), which is more widely recognized (see Fig. 4). In this condition, the pulmonary artery and aorta are transposed. This causes the pulmonary and systemic circulations to function in parallel rather than together. Oxygenated pulmonary venous blood returns to the left atrium and left ventricle but is re-circulated back to the lungs via the pulmonary artery. Deoxygenated systemic venous blood returns to the right atrium and right ventricle, where it is subsequently pumped to the systemic circulation, effectively bypassing the lungs. This parallel circulatory arrangement results in a deficient oxygen supply to the tissues and an excessive right and left ventricular workload. It is incompatible with prolonged survival, unless mixing of oxygenated and deoxygenated blood occurs at some anatomic level (Warnes, 2006).

Embryology

Cardiac development of the fetus occurs between the 3rd and 7th weeks of gestation. As the embryo grows, it divides into 3 layers of cells: the ectoderm, the mesoderm and the endoderm. It is from the mesoderm that the cardiovascular system and the blood and lymph are derived (Sadowski, 2004). The heart and great vessels start forming in the cardiogenic part of the mesoderm, near the cranial end of the embryo. As the embryo grows and thickens, it begins to fold upon itself, both horizontally and vertically (Abdulla et al., 2004; Moore and Persuad, 2008). The heart is moved in front of the foregut and below the oro-pharyngeal membrane. It develops from two longitudinal strands or cords into two endothelial tubes. A single heart tube is then created from these and it elongates, expands and bends to the right (d-looping), forming an S shape. A central swelling, known as the bulbus cordis becomes apparent. This process places the right ventricle on the right and the left ventricle on the left, as they should be. It also brings the atrium to the right and posterior to the ventricles (Moore and