Can fetal echocardiograms reliably predict the need for urgent balloon atrial septostomy in transposition of the great arteries?

**CLINICAL SCENARIO**
A term neonate is admitted to the neonatal unit with an antenatal diagnosis of transposition of the great arteries with intact ventricular septum (TGA/IVS) and a 6 mm patent foramen ovale, identified by fetal echocardiogram at 34 weeks gestational age. The preductal and postductal oxygen saturations ($\text{SpO}_2$) are 45% and 55% in 100% oxygen. Prostaglandin therapy is commenced at 5 ng/kg/min. In view of unresponsive hypoxaemia, the baby is intubated, mechanically ventilated and prostaglandin dose is escalated to 50 ng/kg/min. However, little improvement in oxygen saturations is noted. You then question the need for urgent balloon atrial septotomy (BAS) although fetal echocardiogram suggested adequate mixing at the atrial connection.

**STRUCTURED CLINICAL QUESTION**
In neonates, with an antenatal diagnosis of TGA with IVS or small ventricular septum defect (VSD) (patient), can fetal echocardiogram (intervention) predict the need for urgent BAS (outcome)?

**SEARCH**
Cochrane, Medline, Embase, Cinahl and Maternity and infant care databases were searched on 6 March 2019. The following terms were used: exp ‘INFANT, NEWBORN’/OR (newborn * OR neonates*) AND exp ‘HEART DETECTS, CONGENITAL’/OR (congenital heart AND (defect* OR malformation*)) AND exp ‘ULTRASONOGRAPHY, PRENATAL’/OR ((prenatal OR antenatal) AND (ultrasonography OR ultrasound OR scan)) OR (echocardiogram OR echocardiograph*, OR ‘cardiac echo’) AND exp FETUS/OR exp ‘FETAL RESEARCH’/OR ((fetal OR foetal OR fetus OR fetus OR prenatal OR antenatal) AND ultrasonography OR (echocardiogram OR echocardiograph*, OR ‘cardiac echo’)) AND exp FETUS/OR exp ‘FETAL RESEARCH’/OR ((fetal OR foetal OR fetus OR fetus OR prenatal OR antenatal) AND echocardiog*) AND (‘Balloon atrial septostomy’ OR atrial septostomy), OR Balloon atrial septostomy OR atrioseptostomy). In total, 31 publications were identified, in which 4 studies were relevant on further review. All 4 studies included newborns with an antenatal diagnosis of TGA.

**COMMENTARY**
Congenital heart defects (CHD) are the most common congenital anomaly affecting approximately 8 per 1000 livebirths. Data from the National Institute for Cardiovascular Outcomes Research (NICOR-Congenital) show improving antenatal CHD detection rates across England, with an increase in antenatal diagnoses of TGA/IVS from 27.2% in 2007 to 66.7% in 2016. This correlates with the introduction of routine screening of outflow tracts to the NHS Fetal Anomaly Screening Programme recommended by the National Institute of Health and Clinical Excellence.
TGA accounts for 5%–7% of all CHD.⁸ The survival of neonates with TGA has improved following the adoption of new cardiac surgical techniques and enhanced postoperative management.⁹ Antenatal detection, allowing for planned delivery in experienced tertiary centres and improved early neonatal management have also improved outcomes.¹⁰ ¹¹ Despite these advances, some neonates with known TGA will show profound hypoxaemia after birth, secondary to inadequate circulatory mixing, which can lead to preoperative mortality and morbidity.¹² BAS is an accepted standard treatment. It creates oxygenation of the systemic blood, thereby stabilising patients with an appropriate interatrial communication allowing satisfactory function of FO, and ductal size and shunting pattern. They found a hypermobile septum and reverse diastolic patent ductus arteriosus (DA) had a significant association with urgent BAS. Jouannic et al¹⁵ examined the size of the FO and DA to determine the specificity and sensitivity for urgent BAS. They concluded that a restricted FO and/or constricted DA had a specificity of 84%, to predict the need for urgent BAS, but a low sensitivity of 31%.

In conclusion, all identified studies were retrospective and single-centred with small sample sizes. The individual markers in each study were not universally present in all affected cases, making the interpretation of the results challenging. The findings of each study were variable, with contradictory conclusions.

**Clinical bottom line**

- There are no consistent markers on fetal echocardiography that reliably predict the need for BAS in TGA (grade D).
With the lack of evidence to make a reliable prediction of need for BAS on fetal echocardiograms, neonates with TGA and IVS or small VSD should be delivered in centres with emergency interventional cardiology support, should there be an urgent need for intervention. For those neonates delivered in neonatal units with no interventional cardiology services, there should be an urgency of transferring to a cardiology centre, following commencement of prostaglandin therapy.

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