

## **Outcomes and Risk Stratification of Tricuspid Atresia in Scotland: A 15 year Retrospective Review**

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### **Introduction**

Tricuspid atresia can occur in isolation or in association with other congenital cardiac abnormalities, for example: aortic arch abnormalities; ventriculo-arterial (VA) discordance; varying degrees of right ventricular hypoplasia; pulmonary stenosis and pulmonary atresia. The presence or absence of associated abnormalities influences initial and subsequent surgical options.

### **Objectives**

To report the associated cardiac lesions in our population with tricuspid atresia and assess whether the presence or absence of such lesions alters the long term prognosis.

### **Methods**

A retrospective analysis of 40 consecutive cases of tricuspid atresia diagnosed in the national congenital cardiac centre from January 1st 2000 – December 31st 2014.

### **Results**

A diagnosis of tricuspid atresia was confirmed in 40 children who were live born during the study period, 21 females and 19 males.

Morphology in association with tricuspid atresia included: 22 (55%) with concordant VA connections; five (12.5%) with discordant VA connections; two (5%) with concordant VA connections and an associated aortic arch abnormality; four (10%) with discordant VA connections and an aortic arch abnormality; three (7.5%) with pulmonary atresia and four (10%) with pulmonary stenosis.

Three patients died within the first month of life without surgical intervention; three died post shunt procedure and one died post pulmonary artery band, secondary to associated ventricular failure. 17 patients required a shunt procedure, of which three (17.6%) died post procedure and prior to Glenn palliation. The presence of pulmonary atresia in association with tricuspid atresia resulted in 100% mortality in the 1st year of life. 33 (82.5%) patients survived to Glenn palliation.

There was only one (2.5%) reported case of plastic bronchitis and no reported cases of protein losing enteropathy (PLE) or heart transplantation. There were no deaths after 9 months of age and 33 children are alive to date. Survival to 5 and 10 years was 81%.

### **Conclusions**

Survival in the Scottish population is similar to international published rates of survival following palliation of tricuspid atresia. The presence of associated cardiac lesions allows paediatric cardiologists to risk stratify tricuspid atresia at diagnosis, predicting the initial surgical pathways and ultimately provide parents with a more accurate long term outlook.