

# Summary of the Tricuspid Atresia (TA) Analysis

Dear Family,

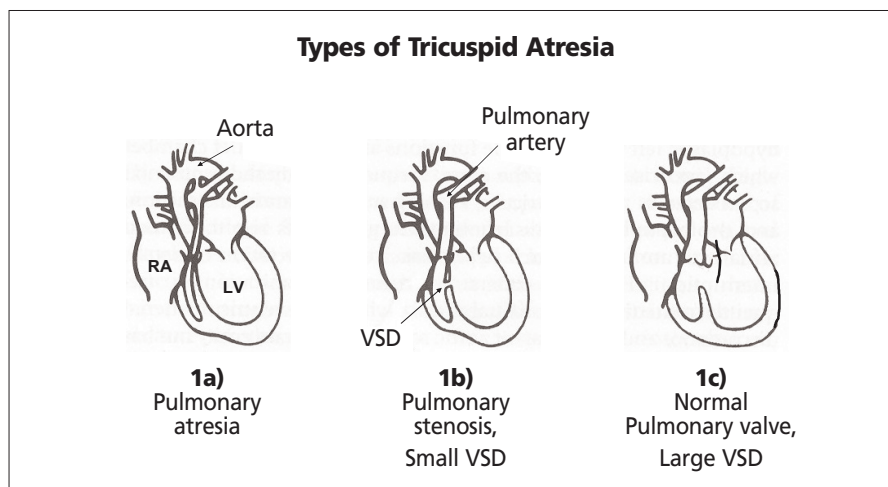
We wish to thank you for your continuing participation in the Congenital Heart Surgeons Society (CHSS) study following children with tricuspid atresia, which began in 1999.

The purpose of this letter is to provide participating families with a summary of results from the recent analysis that was presented at the annual meeting of the American Association of Thoracic Surgery in San Francisco on April 12, 2005. The results have been accepted for publication in the *Journal of Thoracic and Cardiovascular Surgery*, and will likely be published in that journal before the end of this year.

## Tricuspid Atresia Background

Tricuspid atresia is a rare type of congenital heart disease, affecting approximately 2 per cent of all babies born with a heart defect. The tricuspid valve normally located between the collection chamber on the right side of the heart (right atrium) and the right-sided pumping chamber (right ventricle) fails to develop. Therefore, blood that returns from the body to the right atrium cannot directly enter the right ventricle, and must pass through a hole in the atrial septum (atrial septal defect) into the left side of the heart.

There are several different forms of TA that affect the symptoms and course of treatment in any given patient, as shown in the figure below.



*Legend: RA=right atrium; LV=left ventricle; VSD=ventricular septal defect*

There may be complete blockage, (atresia), of the valve (pulmonary valve) between the right ventricle and the artery to the lungs (pulmonary artery). This is referred to as Type Ia. There may be a hole in the ventricular septum, called a ventricular septal defect (VSD) that can be very small (Type Ib) or quite large (Type Ic).

## Different Treatments for Tricuspid Atresia:

Depending on the type of tricuspid atresia, children are treated using an approach that is usually made up of two or three stages. The first stage may be either a shunt or band, the second stage is known as a Glenn operation, and the third stage is known as a Fontan operation.

The aim of this CHSS study is to provide pediatric cardiologists and cardiac surgeons with useful information allowing improved decision-making and better outcomes for children.

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## The CHSS Study

This is an ongoing study, but the initial analysis includes newborns enrolled between 1999 and 2004. During this time, 150 babies who were cared for at 26 participating hospitals were enrolled. The objectives of this study were to determine the number of children reaching the different treatment stages including the second stage (Glenn operation), and the third stage (Fontan operation). We also wanted to determine the number of patients who died, and the reasons for their death.

### A brief summary of the results

For all 150 children enrolled in the study, the percentage that are alive at 6 months after diagnosis was 96 per cent, and later at 5 years of age, was 86 per cent. Two years after diagnosis, 90 per cent of children had the Glenn operation, and by 3 years of age 75 per cent had a Fontan operation. Only 5 per cent of children died before reaching either the Glenn or Fontan operation.

We discovered some important factors that affect which state children are more likely to reach. Factors which increased the risk of a worsened outcome included the function of the left-sided heart valve (mitral valve) that directs blood from the left-sided collection chamber (left atrium) to the left-sided pumping chamber (left ventricle). Children with an incompetent, or leaky, mitral valve therefore require closer follow-up than those children with normal mitral valves.

Children with more normal anatomy whose "shunts" were able to be constructed from a particular blood vessel also had improved outcomes. Factors which increased the chance of reaching the Glenn operation included older patient age at surgery and using a smaller "shunt" size.

The important message from our study is that outcomes of children with TA can be improved by using identified anatomic factors to help guide the type of surgical procedure performed. This strategy may be more effective and result in a lower risk of death and a greater number of children reaching the second or third stage at an earlier age.

We thank you for your continued participation in this CHSS study of children with TA. The information gathered, analyzed, and published by the CHSS is read by pediatric cardiac care specialists around the world. It is your participation that makes these important studies possible and helps us improve the care for children with congenital heart disease.

*Sincerely,*

*William G. Williams, MD  
Christopher A. Caldarone, MD  
Geraldine Cullen-Dean, RN, MN  
Olga Levesque, B.A.  
Haddas Grosbein, B.Sc.*

*Brian W. McCrindle, MD  
Tara Karamlou, MD  
Sally Cai, M.Sc.  
Candice Cumberbach*

***The CHSS Data Center welcomes any questions, comments or suggestions from the participating families. For further information and a full-version of the published manuscript, please visit our Web site at [www.chssdc.org](http://www.chssdc.org) or contact us at:***

### **CHSS DATA CENTER**

Hospital for Sick Children

Room 4433, Gerrard Wing, 555 University Ave., Toronto, ON CANADA M5G 1X8

Telephone: 416-813-8477 Fax: 416-813-8776

**TOLL FREE: 1-866-477-CHSS (2477)**