CHSS Data Center
CONGENITAL HEART SURGEONS' SOCIETY

Summary of the Pulmonary Atresia Intact Ventricular Septum (PAIVS) Analysis

July 2003

Dear family,

We wish to thank you for your continuing participation in the Congenital Heart Surgeons Society (CHSS) study that follows children with pulmonary atresia - intact ventricular septum (PAIVS). The study began in 1987 when the CHSS was based at the University of Alabama, Birmingham. In 1998 the CHSS Data Center moved to the University of Toronto at the Hospital for Sick Children. We continue to follow all children enrolled in each of eight CHSS studies, including the children with PAIVS. The purpose of this letter is to provide participating families with a summary of results from the most recent analysis that was presented at the annual meeting of the American Association for Thoracic Surgery in Boston on May 6, 2003. The results have also been submitted for publication in the Journal of Thoracic and Cardiovascular Surgery, and if accepted, will likely be in print by the summer of 2004.

PAIVS Background.

PAIVS is a birth defect of the heart involving total blockage of the valve (pulmonary valve atresia - the "PA" in PAIVS) between the right-sided pumping chamber (or right ventricle) and the artery to the lungs (pulmonary artery). The blocked pulmonary valve prevents blood flow to the lungs. Therefore, blood flow through the right side of the heart is severely reduced, usually resulting in abnormally small right sided heart structures including the right ventricle and inflow (tricuspid) valve. The tricuspid valve normally prevents blood from flowing backwards from the right ventricle to the right atrium. The wall, or septum, between the right and left ventricles is intact (the "IVS" in PAIVS) - *ie* there is no hole between the ventricles in PAIVS. The coronary arteries, which supply blood flow to the heart muscle, may be abnormal in PAIVS. These many factors result in complex decisions in caring for children with PAIVS. The aim of the CHSS study is to provide pediatric cardiologists and cardiac surgeons with useful information allowing improved decision making and better outcomes for children.

CHSS Study.

The enrollment period for the PAIVS study was between 1987 and 1997. During this time, 408 children who were cared for at one of 33 participating hospitals were enrolled. To be eligible for participation, the babies had to be admitted to a CHSS hospital within the first 30 days of life. The purpose of the study is to determine the percentage of children reaching one of the 6 possible outcomes. We also sought to identify the factors that influence each of the 6 outcomes. This information should allow us to improve results of treating children with PAIVS.

The outcomes and a brief description of each are listed below:

- **2-ventricle repair:** an operation or series of operations that results in two ventricles - one (right) pumping blood to the lungs and one (left) pumping blood from the lungs to the body. The operation(s) corrects the blocked pulmonary valve to allow the right ventricle to pump blood to the lungs. The circulation in this type of repair most closely resembles "normal" circulation.
- **1.5-ventricle repair:** an operation or series of operations as described for 2-ventricle repair with the exception that blood returning to the heart from the upper portion of the body is surgically redirected into the artery to the lungs (cavopulmonary or "Glenn" shunt), instead of going through the right ventricle. Surgeons perform this type of repair when the right heart is inadequate to accept all of the blood flow as required of a 2-ventricle repair.

- **Fontan operation:** a operation that results in all of the blood returning from the body going directly to the lungs without being pumped by the right ventricle. The Fontan operation is usually done several months after a cavopulmonary, or "Glenn", shunt in which blood returning from the upper body is returned directly into the artery to the lungs.
- **Heart transplant:** an operation in which the heart with PAIVS is removed and replaced with a donor heart to create a "normal" circulation.
- **Death before reaching a definitive repair:** children who die before one of the above operations are also considered as one of the 6 outcomes. It is obviously one to be avoided. Identifying the factors that increase or decrease the risk of death are especially important.
- **Alive without reaching a definitive repair:** children who are alive but have not yet reached one of the above defined outcomes.

A brief summary of the results of the study follows. For all 408 children enrolled in the study, the number who are alive at 1 month after hospital admission is 77% and later at 5 years and 15 years is 60% and 58% respectively. Importantly, the risk of death dropped during the 10-year study period. Survival at 5 years improved to 80% by 1997.

By 15 years after first hospital admission 33% of the children had 2-ventricle repairs, 20% had a Fontan operation, 5% had a 1.5 ventricle repair, and 2% had a heart transplant. 38% died before reaching a repair. Only a small number of children (2%) are living without a repair.

We discovered some factors that determine which outcome children are most likely to have. One important factor is the size of the right sided heart structures - the tricuspid valve and right ventricle. Children with normal or near-normal right heart size are more likely to have a 2-ventricle repair, while those with smaller, more severely malformed right sided structures are more likely to have Fontan operation or to die before reaching a repair. Other important factors that determine outcomes include degree of abnormal coronary arterial circulation, lower birth weight, and how well the tricuspid valve functions.

The study shows that outcomes of children with PAIVS can be improved by using the identified anatomic factors to decide which type of surgical repair is most appropriate for each child. This strategy results in a lower risk of death and a greater number of children reaching a repair and at an earlier age.

We thank you for your continued participation in this CHSS study of children with PAIVS. The information gathered, analysed, 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. The CHSS Data Center welcomes any comments or suggestions from the participating families. For further information, please visit our website (**www.chssdc.org**).

Sincerely,

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