

Summary of the Interrupted Aortic Arch (IAA) Analysis A Congenital Heart Surgeons Society Study

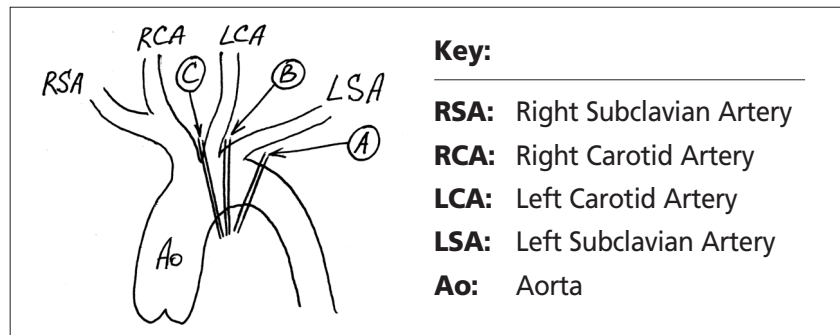
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Dear Family,

We wish to thank you for your continuing participation in the Congenital Heart Surgeons Society (CHSS) study that follows children with Interrupted Aortic Arch (IAA). 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. The CHSS consists of 70 surgeons from 40 institutions in 4 countries (USA, Canada, Brazil & Argentina). We continue to follow all children enrolled in each of eight CHSS studies, including the children with IAA. 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 Toronto on April 26th, 2004. The results have also been accepted for publication in the Journal of Thoracic and Cardiovascular Surgery, and will likely be in print by the summer of 2005.

IAA Background:

IAA is a birth defect of the heart in which a portion of the major artery arising from the heart (aorta) is absent. There are three types of Interrupted Aortic Arch, and they are classified according to the site of interruption.



Type A: Most of the aortic arch is normal but the interruption occurs just beyond the branch that goes to the left arm (left subclavian artery). 29% of the infants with Interrupted Aortic Arch have Type A.

Type B: The interruption occurs between the left carotid artery (one of the arteries to the head) and the left subclavian artery. Type B is the most common form of Interrupted Aortic Arch. It accounts for about 70% of our study children.

Type C: The interruption occurs between the innominate artery (artery to the right arm and head) and the left carotid artery. Type C is the least common form of Interrupted Aortic Arch, accounting for only 1% of the children in our study.

Interrupted Aortic Arch develops as a result of abnormal formation of the aortic arch system during the fifth to seventh week of fetal development. This defect is almost always associated with a large hole between the two pumping chambers, called a ventricular septal defect (VSD). The goal of surgery is to join the aortic arch together and to close the ventricular septal defect and repair other associated heart defects. Surgery is usually performed in the first week of life after the infant is stabilized. Complications after IAA repair may include narrowing at the aortic repair site or within the heart.

CHSS Study.

The enrollment period for the IAA study was between 1987 and 1997. During this time, 472 newborns 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 those factors that affect outcomes of these babies. This information should allow us to improve future results of treating babies born with IAA.

For all 472 children enrolled in the study, 83% were alive 6 months later, 70% survived to age 5 years and 62% to age 15 years. Importantly, the survival rate improved during the 10 year study period.

We discovered some important factors that may cause a poor outcome. These factors are lower birth weight, younger age at study entry, type B IAA and the presence of any other major heart problems besides IAA.

Sixteen years after repair of IAA, 66% of the children are alive and 28% have had another procedure because a narrowing that developed at the arch repair. Arch procedures (either surgery or balloon catheter dilation) were more likely to occur in children who required a repair of a major associated heart defect called truncus arteriosus, or had a repair with a fabric material called Gortex as part of the arch repair.

Our analysis shows that the best method of IAA repair is to directly sew the ends of the aorta together and to make this connection bigger with a patch that is not Gortex. This type of repair reduces the chance of death and reduces the risk of later narrowing of the IAA repair.

Sometimes children with IAA have narrowing that obstructs blood flow from the outlet of the left side of the heart. These children with outlet obstruction had poorer survival.

If all of the risk factors we identified in this study could be eliminated, the 16 year survival would be 93%, with a 24% chance of needing a repeat procedure for narrowing at the IAA repair site, and a 16% chance of needing an initial or repeat procedure to relieve obstruction of blood flow from the left side of the heart.

ACKNOWLEDGEMENT

We thank you for your important and continued participation in this CHSS study of children with IAA. 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 understanding and 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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