Professor Richard A. Jonas: exploring ways to improve brain development in children with congenital heart disease

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Introduction

Professor Richard A. Jonas joined the staff of Children's National Medical Center in September 2004 as the Chief of Cardiovascular Surgery, Co-Director of the Children's Heart Institute and Cohen Funger Professor of Cardiac Surgery. He undertook his general surgical training at the Royal Melbourne Hospital in Melbourne Australia and subsequently his cardiothoracic surgical training at Royal Children's Hospital in Melbourne and Green Lane Hospital in Auckland New Zealand. After fellowships at the Brigham and Women's Hospital and Children's Hospital Boston he was appointed to the Department of Surgery at Harvard Medical School in 1984. In 1994 Professor Jonas was appointed to the William E. Ladd Chair of Surgery at Harvard Medical School and became the Cardiovascular Surgeon in Chief at Children's Hospital Boston.

Professor Jonas has an active clinical practice in congenital cardiac surgery. He also maintains his own National Institutes of Health (NIH) supported laboratory research program in addition to his administrative responsibilities. He is the author of over 360 peer-reviewed publications and 2 textbooks.

In addition to his responsibilities at Children's Hospital, Professor Jonas is a consultant to the FDA and has been a regular member of the CICS study group of the NIH. He has worked with Project Hope for more than 25 years in establishing pediatric heart surgery in Shanghai, China as well as helping with the design and development of the Shanghai Children's Medical Center.

In June 2005 Professor Jonas was the President of the American Association for Thoracic Surgery and in October 2009 was President of the Congenital Heart Surgeon's Society of North America. He is currently vice-President of the World Society for Pediatric and Congenital Heart Surgery.

As one of the renowned pediatric cardiac surgeon, Professor Richard A. Jonas (*Figure 1*) has been invited to give three important lectures on the 23rd Annual Meeting of the Asian Society for Cardiovascular and Thoracic Surgery



Figure 1 Professor Richard A. Jonas, Chief, Cardiac Surgery; Co-Director, Children's National Heart Institute, Children's National Health System, Washington, DC, USA.

(ASCVTS) in Hong Kong in 2015. With this great chance, we are honored to have an interview with Professor Jonas to share his researches and experience in managing congenital heart disease (*Figure 2*).

JTD: According to your decades of clinical and research experience, what is the most common heart disease in children in the past decade? Is its incidence increasing?

Prof. Jonas: About a little less than one percent of the population have congenital heart problems in the world. By far, the commonest problem is a hole in the heart, what we call ventricular septal defect (VSD). Patients can have an isolated VSD (just the VSD by itself) or VSD in combination with many other problems. But VSD is as common as any problem we deal with surgically. Its

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Figure 2 Professor Richard A. Jonas and JTD's Science Editor Chao-Xiu (Melanie) He, were on the conference site of the 23rd Annual Meeting of the Asian Society for Cardiovascular and Thoracic Surgery (ASCVTS) in Hong Kong.

incidence appears to be very stable, with better diagnosis in developing countries like China, but actually the incidence is quite stable.

JTD: As you have had a specific lecture about surgical option for cavopulmonary connections, how do you look at the two options: extra cardiac vs. lateral tunnel?

Prof. Jonas: What I explained in my lecture is there has been a big change over the 40 years that people have been doing a Fontan Operation. Forty years ago people did a simple operation; then for the next 10 years, the lateral tunnel operation; then for the next 10 years, the extracardiac conduit. And now, some people including me have changed to what we called intra/extracardiac conduit. It's actually on the front cover of my book. In it, it shows how the intra/extracardiac conduit is used to do operations. I have been using it for about 10 years now.

JTD: In your lecture "optimal timing for elective repair of TOF", you propose to adopt elective repair as early as possible, what are the major reasons for this?

Prof. Jonas: The age people fix heart problems has been coming down. Surgeons used to try to delay the repair operation by doing palliative operations, a simple operation to let the child stay alive but not fix the problem. So the age of repair has been gradually moving down, and we now recommend that tetralogy should be fixed in the first few weeks of life.

JTD: As far as I know, you and your team are devoted to developing safer cardiopulmonary bypass techniques to improve the safety and patient outcomes. What is the major advances in the bypass technique in recent years, and how do they affect the patients outcomes?

Prof. Jonas: The most important change is that the machine has become much smaller. Because it is a pump, the heart and lung machine needs some fluid inside it. If it is big, the fluid has to be a lot of blood from the blood bank, not the baby's blood. So to mix somebody else's blood, maybe blood from four or five people with baby's blood in itself can make the baby sick. Today, the heart and lung machine pump is very tiny, so in some operations there is no need to add any blood. So there is much less bad effect from a blood transfusion. Also the machines, the pumps cause less damage to the blood which used to cause a lot of inflammation, so the baby would feel like having an infection. Now there is much less inflammation, so there is less swelling, fever or other side effects. I think the biggest improvement is to use heart and lung machines that are specifically designed for babies.

JTD: Would you like to share the latest research program that your team is conducting?

Prof. Jonas: The research that we are doing is supported by NIH in the US. We are trying to improve brain development in children who have heart problems. The heart develops very early in the baby's development. When the fetus is just two months' old, the heart is already fully formed. So for seven months, the heart is pumping inside the baby before the baby is born. If you have a congenital heart problem, sometimes you do not get enough blood going to the brain, then you don't get oxygen going to the brain. Then the brain does not develop normally. So we find some kids have problems, mainly in behavior like ADHD attention deficit/hyperactivity disorder and difficulty in concentrating. Sometimes they have difficulty with motor skills like picking up small objects, and sometimes they would have walking imbalance. So we are presently trying to understand better how the congenital heart problems affect the developing brain before birth, and we would be able to either change the circulation before birth or at least treat the brain after birth, to make sure the child has less risk of having behavior problems.

We are now just trying to understand what the problem is by doing MRI scans when baby is born and after the

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surgery. It used to be that the brain problem would get even worse after the surgery because the heart and lung machine would affect brain development even more. So they would harm the brain. But today most of those problems caused by the heart and lung machine are very minor. Children with simple heart problems do not usually have brain problems. It is very complicated heart problems that are associated with a lot of behavior problems so that some kids can not go to school and college. They can do many regular things, but they just have some problems in behavior.

JTD: As you are an experienced pediatric cardiac surgeon and have trained a large number of young surgeons, do you have anything to share with those who would like to become an excellent pediatric cardiac surgeon?

Prof. Jonas: Chapter 2 of my book *Comprehensive Surgical Management of Congenital Heart Disease* is called "Becoming a Congenital Heart Surgeon", and it explains what is needed to become a pediatric heart surgeon. Because if you operate on a child who dies, the family can be very sad, sometimes the family can be very angry at you. So this is a very big challenge in caring for children who have heart problems. The chapter explains before you choose to become a

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congenital heart surgeon, you need to think carefully and you have the right personality to be able to manage the emotional challenges and physical challenges. And you have to understand some very complicated things. Today we have many surgeons come from China, India and from other countries. It is quite difficult to get a training position in the United States. You need to do a lot of exams, you need to get visas, you need to get medical license, you need to get medical privileges, so many things. So chapter 2 explains what you need to do to become a congenital surgeon.

JTD: Thank you very much for your generosity of time for the interview!

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None.

Footnote

Conflicts of Interest: The author has no conflicts of interest to declare.

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