THE BAIRD INSTITUTE
Applied heart & lung surgical research

'Our mother's death was not only deeply distressing but utterly unexpected. She'd lived a virtually illness-free life and looked a picture of health until the day of her death'

Dear Friends

Shortly you will read the amazing story of Jake Rance and his family and their experience with Familial Thoracic Aortic Aneurysm and Dissection, commonly known as TAAD. Stories like Jake's are the most personal and compelling way of highlighting the importance of our work. While nothing can change the loss of Jake's mother, Jeannie, his story reflects the seriousness of aortic aneurysm and the critical importance of early detection which can lead directly to enhancing the results of surgical outcomes. The enormous importance of research in terms of understanding the genetic nature of aortic aneurysm and aortic disease, enables us to not only provide enhanced patient care, it also increases the likelihood of avoiding the passage of this disease to the next generation.

Established in 2001, in honour of the late Professor Douglas Baird, The Baird Institute is the only group in Australia to focus entirely on clinical and surgical cardiothoracic conditions. The Baird Institute relentlessly pursues innovation – not for innovation alone, but for the potential benefits it can bring to the thousands of women, men and children, who every year, are diagnosed with some form of cardiac condition. Our constant focus is to improve cardiac surgical techniques. As a priority, we are working to advance the field of minimally invasive cardiac surgery and the application of modern medical technology, to support the development of new surgical techniques that improve patient outcomes.

Jake in Royal Prince Alfred Hospital with brother Tim

The Baird Institute receives no government funding.
To do our work we rely very much on the generosity of our supporters, many of whom are current or former patients of the surgeons associated with The Baird Institute. Your generous gift is vital for the continuation of our research. Our goal is to give people a second chance to live a full and healthy life. Few gifts are more precious than that.

In Australia diseases of the heart, lung and blood vessels kill more people than any other disease. These diseases can affect people at any age at any time – even babies and young children. Your gift today will enable us to fund more research that will save lives.

Wishing you good health and happiness.

Professor Paul Bannon PhD MBBS FRACS
Chair

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‘THE HEART OF OUR FAMILY’
BY JAKE RANCE

‘Our mother’s death was not only deeply distressing but utterly unexpected. She’d lived a virtually illness-free life and looked a picture of health until the day of her death’

Ma died from a dissecting aortic aneurysm — or more precisely, a dissection of the aortic root, this is also referred to as **TAAD – Familial Thoracic Aortic Aneurysm and Dissection**. Six years later, while on holiday in Australia, our maternal aunt was admitted to hospital with mildly distressing symptoms, none of them seemingly cardiac related. Some hours after admission, Val lost consciousness and two days later she died. The cause, once again, was TAAD.

Jake and his adored mother, Jeannie

For my two younger brothers and I, it became increasingly clear that losing two adored members of our family had not only left a gaping emotional hole, but a genetic legacy. Following our aunt’s death, the intensive care consultant who had cared for her with such compassion and commitment, now strongly counselled us to pursue our own cardiac investigations. Taking his advice, I found myself several months later in the consulting rooms of the inimitable Professor Richmond Jeremy, a specialist in aortic disease. Early the next year our middle brother Jason followed suit.

Scans of both our aortic roots revealed a degree of dilation nominally outside the bounds of ‘normal’, intimating a possible weakness in the connective tissue but not definitively so. Meanwhile our young brother Tim, a doctor in Brisbane, had painstakingly tracked down both Jeannie and Val’s medical histories. In both cases, neither of their aortas had been overly dilated either. We were not sure how to proceed.

Sensing our concerns were best answered by someone with extensive and literal ‘hands on’ experience, Richmond referred us to Professor Paul Bannon, a cardiothoracic surgeon with impeccable credentials and particular expertise in aortic root surgery. Furnished with our various scans and our burgeoning cardiac pedigree, Jason and I met with Paul to discuss our options. During our conversation it became clear that surgery was less a matter of if than when and by the end of that first meeting potential dates for surgery had been discussed.

It was decided that as I was the eldest it was only fair that I go first! My operation was set for 6 January 2016, only a few months away, and Jason’s for just six weeks later. Ultimately both of us found that making the decision to operate was not a difficult one: if intervention was inevitable at some point then better now while we were both otherwise healthy and young(ish)! For although we were both symptom free we had also witnessed firsthand the catastrophic outcome once dissection occurs. And besides, both our mother and aunt had also been symptom free until their final hours.

The day of my surgery, RPA was holiday quiet. I felt relaxed and optimistic. I was ready! Following a 6-hour long operation, I woke up in intensive care a mess of catheters, tubes, drains, and drips. I was also awash with fentanyl, an analgesic reputedly a hundred times stronger than morphine. And I needed it! Coughing, sitting up, walking … these were all activities that staff gently cajoled and supported me through; simple tasks, seemingly insurmountable at times. Three days in intensive care was followed by four on a post-operative ward. Seven days after surgery I was free to go.

Some six weeks later it was Jason's turn. He too was under the excellent care of Professors Jeremy and Bannon. And once again brother Tim flew down to be with us. Visiting Jason in RPA's intensive care and then later in the same post-operative ward — 'my' ward — felt surreal. In time Jason felt robust enough to join me twice a week at cardiac rehab, a fabulous service offered for free by most large public hospitals. I cannot over-state not only the physical but the psychological merit of sharing one's recovery with others in the same post-operative boat.

Nearly a full 12 weeks after surgery I returned to work. Now, over a year since my surgery, my once impressive scar has faded, and so too has that sense of physical and emotional vulnerability that had once been my constant companion. Jason too has made a full recovery. We have both been the beneficiaries of not only the excellent care of two fine doctors, but blessed to live in a part of the world where a medical system affords us (all) access to their expertise.

Nonetheless, elements of our family's story remain unresolved. My genetic testing revealed nothing that might assist brother Tim with his decision about future surgery, nor inform potential courses of action for Arish, my cherished 10-year-old, or his five cousins.

The imperative to ensure ongoing research, alongside innovative practice, is clear.

In the meantime, for Jason and I, if the deaths of our darling mother and beloved aunt have a silver lining, it's that our own prognoses are now shiny bright!

Please support The Baird Institute to continue its life-saving research.

Jake (left) and Jason Rance

Jake Rance

TAAD – FAMILIAL THORACIC AORTIC ANEURYSM AND DISSECTION

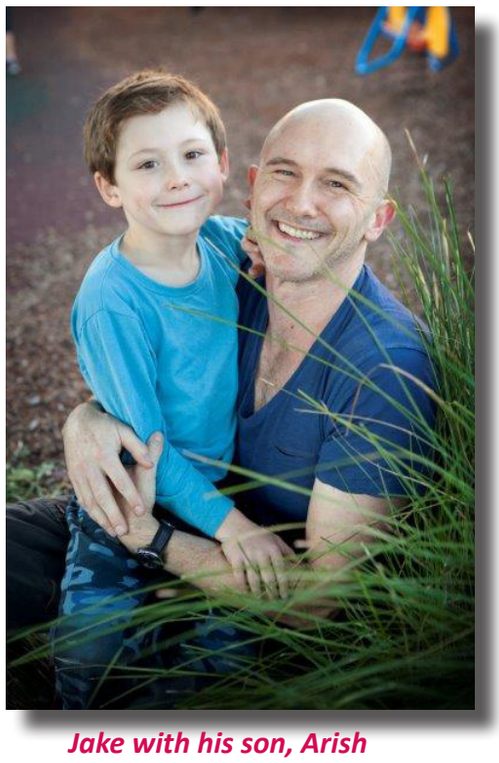
TAAD is a disorder that involves problems with the aorta, which is the large blood vessel that distributes blood from the heart to the rest of the body. Familial TAAD affects the upper part of the aorta, near the heart. This part of the aorta is called the thoracic aorta because it is located in the chest (thorax). In familial TAAD, the aorta can become weakened and stretched (aortic dilation), causing a bulge in the blood vessel wall (an aneurysm). Stretching of the aorta may also lead to a sudden tearing of the layers in the aortic wall (aortic dissection), allowing blood to flow abnormally between the layers.

TAAD is rapidly becoming one of the most common but silent killers in Western society. Forty percent of people with ascending aortic dissections die instantly and according to the American Heart Association, the risk of death increases 1% every hour that diagnosis and surgery are delayed. Up to 90% of patients can be saved with emergency surgery, if they can get to a hospital that provides advanced cardiac surgery.

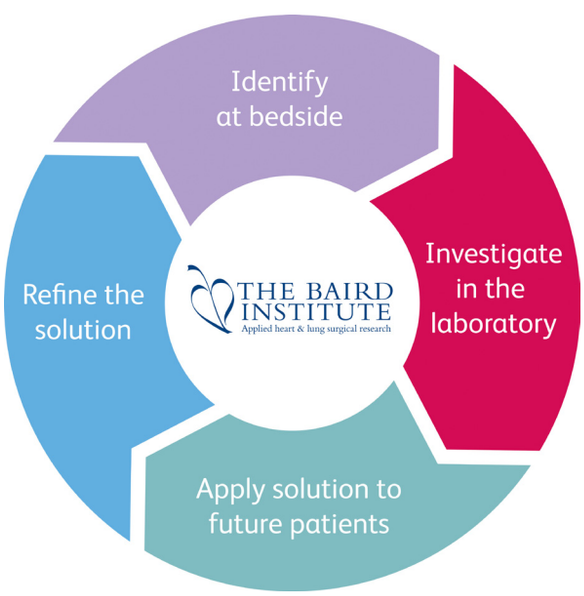
OUR RESEARCH INTO AORTIC DISEASE

Our research program is giving people like Jake and his brothers a second chance at life. The Baird Institute is continuing its involvement in and initiation of research into the prevention and management of aortic disease. Our goal is to determine when surgical intervention is optimal or at the lowest risk for the patient. The Aortic Diseases research group is led by Professors Paul Bannon and Michael Vallely and includes one of our past research fellows, Dr Andrew Sherrah.

Novel Imaging Techniques in Aortic Aneurysm & Dissection: With the Sydney Translational Imaging Lab (at the University of Sydney), new methods and techniques utilising MRI and its ability to assess aortic blood flow over time are being investigated. In patients with both aortic aneurysm and/or long-standing aortic dissection, the relevance and applicability of assessing the derangements in blood flow patterns that can occur is being studied.



Jake with his son, Arish



The late Professor Doug Baird's vision was to identify the problem at the bedside, investigate it in the laboratory and apply the solution in daily practice.

The RPAH/University of Sydney Aortic Tissue Bank: All patients undergoing any aortic repair or replacement at RPAH are being asked to donate their resected tissue to the Aortic Tissue Bank. This is run and sponsored by The Baird Institute. This has allowed researchers to identify proteins and structural derangements in the aortic tissue of large numbers of patients. This will not only allow a better appreciation of the pathological processes associated with aneurysm and dissection, but give new opportunities to investigate therapies to slow or reverse these processes.

The RPAH Patient Database: Another initiative also of the Baird Institute is the collection of data from all patients at Royal Prince Alfred Hospital that allows for the long-term follow-up and assessment of the prosthetic valves and conduits used in the repair and replacement of diseased aorta. This assists in the identification of potential improvements in surgical technique and implants in aortic surgery.

Worldwide Research Collaboration: With the Baird's support, the participation in several international research projects, based in the USA and Europe, has been facilitated. This has included working with the IRAD group, the International Registry of Aortic Dissection.

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The Baird Institute, 305 / 100 Carillon Avenue, Newtown NSW 2042 Tel : 02 9550 2350 ABN 38 096 746 806
info@bairdinstitute.org.au www.bairdinstitute.org.au Facebook.com/bairdinstitute/