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Original Article

The birth of a new era: the introduction of the systemic-to-pulmonary artery shunt for the treatment of cyanotic congenital heart disease*

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Abstract Cardiac surgery was revolutionized on November 29, 1944, when Eileen Saxon underwent the first systemic-to-pulmonary artery shunt at Johns Hopkins University in Baltimore, Maryland, United States of America. The systemic-to-pulmonary artery shunt was initially developed in the laboratory and then applied to patients through the unique collaboration of Vivien Thomas, Alfred Blalock, and Helen B. Taussig. This innovation was the first operation to successfully treat cyanotic cardiac disease. The history of the first operation to successfully treat cyanotic heart disease is an extraordinary history of courage, innovation, and scientific breakthrough. Just as striking is perhaps the ability of the protagonists of this story to overcome seemingly insurmountable barriers of racial and gender discrimination and revolutionize medicine.

Keywords: Tetralogy of fallot; congenital heart disease; cyanotic congenital heart disease; systemic-to-pulmonary artery shunt

There is nothing more difficult to take in hand, more perilous to conduct, nor uncertain in its success, than to take the lead in the introduction of a new order of things. For the innovator has for enemies all of those who have done well under the old, and lukewarm defenders in all of those who may do well under the new
(Niccolo' Macchiavelli, 1469–1527).

Before 1938, treatment of congenital heart disease was universally considered utopic. Robert E. Gross of Boston was the first to venture in this insidious arena in that year when, against the advice of his Chief, William E. Ladd, he successfully ligated a patent ductus arteriosus.[1] This milestone pertained, however, only to a specific malformation and did not address any other, more complicated, forms of congenital heart disease. As eloquently stated in the manuscript describing the first systemic–pulmonary shunt,[2] "A blue baby with a malformed heart was considered beyond the reach of surgical aid". Children with a multitude of cyanotic cardiac malformations had absolutely no hope for a meaningful survival until 1944. They were most often confined to a ward where the diagnostic armamentarium – history and physical examination, as well as plain radiography and fluoroscopy – was as limited as the therapeutic means of alleviating symptoms – oxygen supplementation and diuretics. This left untreated thousands of children whose sole certain destination was death. Physicians caring for these children had developed substantial knowledge of morphologic features of cyanotic cardiac conditions,[3] most often unfortunately derived from the post-mortem findings of those very same children they had helplessly cared for.

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A video of this presentation can be viewed at the following hyperlink: [http://www.allkids.org/wcpccs].

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Figure 1.
Helen Brooke Taussig (1898–1986), photographed by Yousuf Karsh. (Courtesy of the Johns Hopkins University photographic archives).

Figure 2.
Alfred Blalock (1899–1964), photographed by Yousuf Karsh. (Courtesy of the Johns Hopkins University photographic archives).

The history of the first operation to successfully treat cyanotic heart disease is an extraordinary history of courage, innovation, and scientific breakthrough. Just as striking is perhaps the ability of the protagonists of this story to overcome seemingly insurmountable barriers of racial and gender discrimination. At the time in which this operation took place, our hospitals were as segregated as was American society, and women had limited access to higher education, let alone to key leadership positions in health care. The clinicians mentioned herein were able to establish a model of collaboration that, through astute clinical observation and laboratory investigation, transcended these barriers and made treatment of cyanotic children a reality.

The protagonists

Helen B. Taussig (1898–1986)

Helen Brooke Taussig (Fig 1) originally transferred from her native Boston to Baltimore because The Johns Hopkins University was the first Medical School to enrol women in the medical student body. Harvard Medical School did not grant access to women until 1945. Beyond gender discrimination, Helen Taussig had additional barriers to overcome: she was severely dyslexic and profoundly deaf. She mastered cardiac auscultation with the aid of a stethoscope amplifier that she would bring on ward rounds; this eventually enabled her to correlate auscultatory findings with the clinical and pathological diagnosis of many forms of cyanotic heart disease.[2] Importantly, she astutely observed the increase in cyanosis in infants and children that followed clinical closure of the patent ductus arteriosus and inferred that, if one were able to provide an additional source of pulmonary blood flow, cyanosis would improve. In 1939, she travelled to Boston, where she proposed such concept to Robert Gross. In her own words, Robert Gross, ironically a native of Baltimore, was, in 1939, "in the first flash of success of brilliance of closing a duct and thought that there could be nothing more stupid than creating an artificial duct".[4] Taussig was deeply disappointed by such reluctance to help her in bringing her thoughts to fruition. When she consulted with her father – who was on the Faculty of the Harvard School of Economics – about the possibility of remaining in Boston to continue her quest, his advice was "don't go where you are tolerated… go where you are wanted".[4] Helen Taussig returned to Baltimore to focus on congenital heart disease. Alfred Blalock had just arrived from Vanderbilt as the new Chief of Surgery (1941) and had never even ligated a ductus arteriosus up to that point.

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Following the first systemic-to-pulmonary shunt, Helen Taussig remained a pivotal figure in the advancement of the field of paediatric cardiology in the United States. She was awarded the Lasker award (1954) and the Presidential Medal of Freedom (1964), and was elected in 1965 as the first female president of the American Heart Association.

Alfred Blalock (1899–1964)

Alfred Blalock (Fig 2) arrived in Baltimore at the age of 42, as the new Chief of the Department of Surgery. He graduated Medical School at the Johns Hopkins University (1922), but was one of the early casualties of the rigorous residency training programme, which had roots in the tenure of William Halsted.[5,6] After being dismissed from the training programme, he transitioned to training in urology and otolaryngology. Although a mediocre medical student by grades, Alfred Blalock had unquestionable intellectual curiosity, which he readily applied as a resident. He not only introduced the concept of cross-innervation to aid in the treatment of recurrent laryngeal nerve transection after thyroid surgery, but was also the first to report improvement of myasthenia gravis following thymectomy.[6] As Helen Taussig, Alfred Blalock had himself overcome physical barriers, having survived a severe form of pulmonary tuberculosis during residency training. When he returned to Hopkins in 1941, he was acclaimed as a brilliant investigator, with his pioneering work on haemorrhagic shock; this work served as the cornerstone of the rapidly adopted practice of blood transfusion during and after the second-world conflict.[7]

Alfred Blalock was approached by Helen Taussig to help her find the solution to two clinical problems: aortic coarctation and the treatment of cyanosis in patients with congenital heart disease. The two clinical questions interplayed in the eventual solution of the latter. Blalock had devised a procedure for the treatment of coarctation – the Blalock–Park operation – which entailed turning down the subclavian artery to bypass the obstruction.[8] The procedure was investigated in the animal laboratory and was abandoned because of the unacceptable rate of paraplegia in animals that had not developed collateralisation, and whose cardiac output had to be carried through the relatively small subclavian artery. The procedure, however, demonstrated that the subclavian artery could be mobilised and diverted, and that the limb of the animal would not suffer ischaemic damage by sacrificing its main source of blood flow. That same strategy – the anastomosis between subclavian and pulmonary arteries – had been used by Blalock while at Vanderbilt to induce unilateral pulmonary

Figure 3.
Anna, the first long-term survivor of the Blalock–Taussig shunt. Anna is pictured with Michael Shirmer, the 29th "blue baby" to undergo the operation (Time Magazine, 14 March, 1949). Mr Shirmer went on to complete repair and is alive at 73 years of age. Anna in the only animal to have rightfully deserved a portrait to be displayed at the Johns Hopkins Hospital.
(Courtesy of Time Magazine and with permission from Mr Michael Shirmer).

hypertension. Blalock was unsuccessful in creating a model of pulmonary hypertension, but did induce cyanosis and had now a means of augmenting pulmonary arterial blood flow. The concept was taken once again to the animal laboratory, where successful long-term survival could finally be obtained in the canine model (Fig 3).

Vivien Theodore Thomas (1910–1985)

Vivien Thomas (Fig 4) was, initially, the unsung protagonist of the development of the treatment of cyanotic congenital heart disease. Thomas was the grandson of a slave, and could never go beyond high school because of severe financial difficulties that were only compounded by the Great Depression. He was eventually hired as a laboratory technician by Alfred Blalock at Vanderbilt University, where he personally executed many of the procedures that were to later contribute to the clinical application of the Blalock–Taussig shunt. Vivien Thomas worked as a carpenter and had, reportedly, amazing technical dexterity that he brought to these early experiments. When he arrived in Baltimore, he was hired as a janitor. In 1941, no man of colour could in fact be

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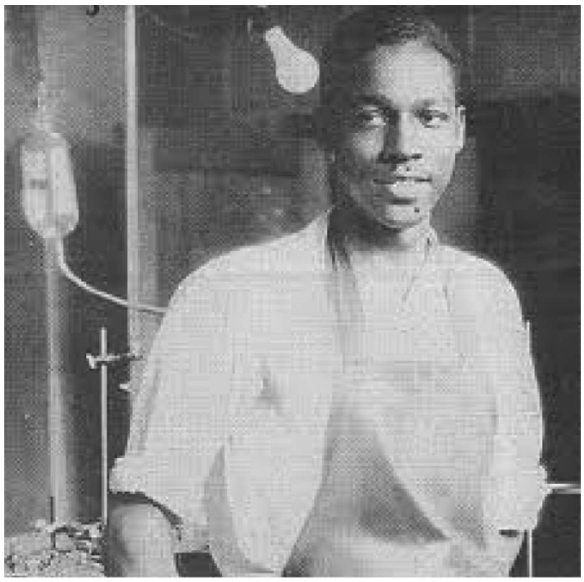

Figure 4.
Vivien Thomas (1910–1985) as a young laboratory technician at Vanderbilt University.
(Courtesy of the Johns Hopkins University photographic archives).

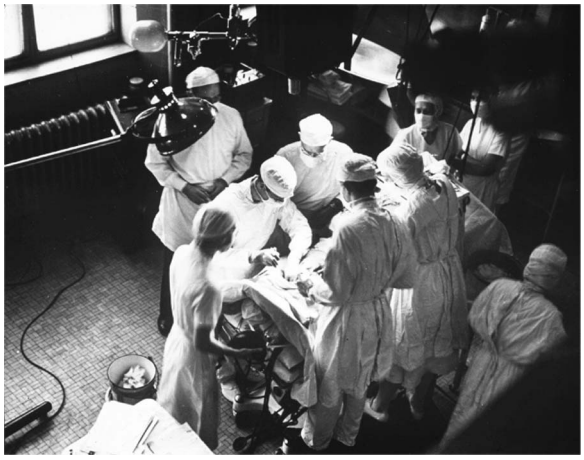

Figure 5.
The "blue baby" operation. Alfred Blalock is seen on the left side of the table. To his left is William P. Longmire (Chief Resident) and, opposite, Denton Cooley (Assistant Resident). Standing behind Alfred Blalock is Vivien Thomas, the only Afro-American who could enter the operating theatre.
(Courtesy of the Johns Hopkins University photographic archives).

appointed to a higher position at Johns Hopkins. In spite of these utter racial barriers, he actively participated in these early operations (Fig 5). For many years, Thomas was tragically unrecognised as a true protagonist in the development of the Blalock–Taussig shunt. He was later nominated as Director of the Surgical Laboratory at Johns Hopkins and, under his tenure, a generation of leaders in cardiothoracic surgery were trained. Some

Figure 6.
Vivien Thomas is finally recognised with a Honoris Cause *Doctorate in 1976. To his left are Dr Helen Taussig and Dr Steven Muller (President of Johns Hopkins University).*
(Courtesy of the Johns Hopkins University photographic archives).

of these went on to play a key role in the early development of closed and, eventually, open heart surgery in children; among these were Denton Cooley, David Sabiston, Henry T. Bahnson, William Muller, and Dwight McGoon. In 1976, Thomas was awarded an honorary Doctorate by the Johns Hopkins University (Fig 6).

Eileen Saxon (1943–1945)

The first patient to undergo the "blue baby" operation was Eileen Saxon (Fig 7), born with tetralogy of Fallot at the prohibitive weight of 1.0 kg. By the time she was 14 months, her weight was 4.0 kg and she was experiencing hypercyanotic spells several times daily. William P. Longmire, the then Chief Surgical Resident and perhaps most accurate Blalock Biographer,[6] vividly depicted Eileen's appearance in the diary he kept as a Resident.

> … On evening rounds, we arrived at the crib of this fifteen-month old baby… I was immediately astounded by the deep cyanotic appearance of the child, much more cyanotic than any patient I had ever seen before: the lips were a deep, dark blue… The face was suffused with dilated veins, the conjunctiva almost purple
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> From: *Alfred Blalock: His Life and Times.*

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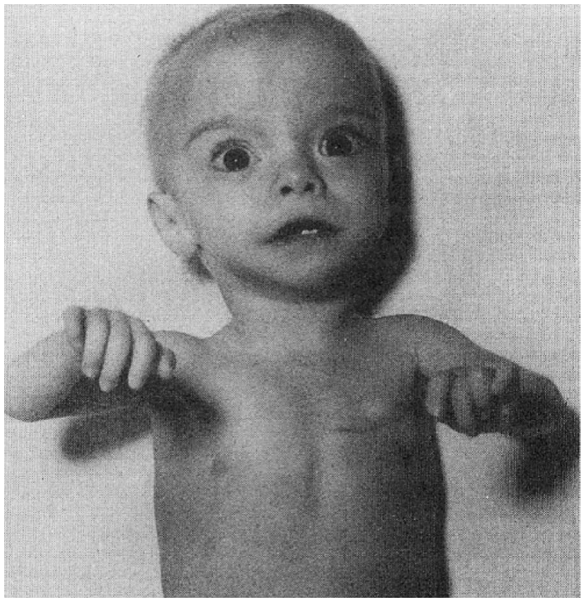

Figure 7.
Eileen Saxon (1943–1945), the first baby to undergo a systemic–pulmonary shunt. Note the left-sided scar, ipsilateral to the aortic arch.
(Courtesy of the Johns Hopkins University photographic archives).

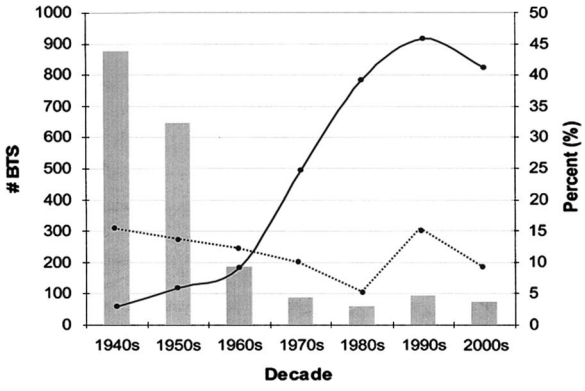

Figure 8.
Evolution of the Blalock–Taussig shunt at the Johns Hopkins Hospital over six decades. Number of shunts (#BTS) per decade, percentage operative mortality (dotted line) and percentage of single ventricle (solid line). The resurge in mortality in the 1990s is likely secondary to the introduction of the Blalock–Taussig shunt as integral part of the Norwood procedure.[10]
(Reproduced with permission from The Annals of Thoracic Surgery).

Eileen Aileen Saxon underwent the operation successfully, but succumbed several months later after re-operation to overcome recurrent stenosis.

The operation

The operation took place on 29 November, 1944 (Fig 5). In the first procedures, an anterior, high thoracotomy was performed on the side ipsilateral to the aortic arch. This approach was rapidly recognised as unfavourable, and was changed to a contralateral, posterior thoracotomy as soon as the first right aortic arch was incidentally encountered at surgery. With the latter approach, torsion of the subclavian artery could be avoided. Eileen rapidly decompensated during clamping of the left branch pulmonary artery, and mask ventilation had to be converted emergently to endotracheal intubation with a makeshift urological catheter. At the end of the operation, a child who came into the operating room profoundly cyanotic exited for the very first time alive and pink, an achievement that had a magnitude that might be hard to fully comprehend in this time of technological wonder. The collaboration between surgeon, cardiologist, anesthesiologist, and laboratory technician had produced the first successful treatment of cyanotic congenital heart disease.

The legacy of the first systemic–pulmonary artery shunt

The operation immediately demonstrated that cyanosis could be treated successfully. Erithrocytosis was reversed, and children could increase their level of activity without breathlessness or the need for squatting. This palliation could last for years, and did so for many who eventually underwent complete correction. Over the next decade, patients worldwide were referred to Baltimore for initial palliation.

The shunt has changed over the years in its most commonly utilised configuration and indications. Following the introduction in 1981 of the modified Blalock–Taussig shunt by Professor Marc R. de Leval, MD, FRCS, and colleagues, from The Great Ormond Street Hospital for Children,[9] classic shunts have been almost universally abandoned in favour of the more predictable, flow-regulated interposition graft now typically performed via a median sternotomy. The change in indications that ensued over the six decades following the original procedure is represented in Figure 8. Following an initial surge in procedures performed in children with biventricular heart disease, shunts are now mainly performed as the initial step in the palliation of infants with univentricular cardiac disease characterised by pulmonary hypoperfusion or ductal dependence.

Although the indications, technique, and results have substantially changed over the past six decades, the history of this operation still represents a triumph of determination, clinical acumen, and scientific investigation over seemingly insurmountable social barriers.

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