**Title Page**

**Type of Article: Feature Article Elective Reflection**

# Title of article: BT shunts, Berlin Hearts and Brave Decisions

**Source of submission: Original work**

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**Mini Biography: Nicole is a final year medical student with an interest in medical education, surgery and ethics. She is a keen writer, runner and painter. She enjoys combining her talents to improve the capacity of her peers and patients.**

**Summary: Reflection of a paediatric cardiac surgery elective demonstrating high-level care. Unravelling the broad systems at play that influence clinical outcomes through the lens of history, innovation, and present-day communication strategies.**

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**Abstract**

An elective in paediatric cardiac surgery provides a unique insight into a niche specialty. The high-level care requires multiple systems to align. Within the patient lies the heart, a mechanical and biological system. Any intervention performed on the heart impacts the wider clinical context requiring the treating team to balance technical feasibility with the functional needs of the patient. Risk stratification, collaboration, and prioritising quality of life are the basis of modern day paediatric cardiac surgery. However, the execution of these goals is dependent upon the evolution of knowledge, technology, and communication. In drawing upon historical milestones in the field and current management strategies, the author highlights the complexity and innovation involved in paediatric cardiac surgery.

## Introduction

The Hospital for Sick Children in Toronto Canada is a centre rich in innovation where I was fortunate enough to spend my medical elective observing paediatric cardiac surgery.



To be present for such life-changing procedures provided many opportunities to consider what is required for high-level care.  Upon reflection, many systems are at play. The biological heart as a mechanical system is influenced by surgical intervention, which impacts upon the patient as a functioning being in the context of future capacity, quality of life, and family. Another critical element is information transfer. Communication is needed for the treating team and patient's family to reach a shared understanding of care intent and management. Shared understanding also moves beyond the therapeutic process of the current patient to consider future patients. The evolution of knowledge is ongoing. At any one time, the procedures performed are governed and defined by knowledge, and technology developed and refined overtime from historical practice and research. Indeed, high-level care is multi-faceted and dynamic.

In order for a unified outcome to be achieved, such intricate inter-related networks must synchronise in a syncytium, much like the intrinsic cardiac muscle fibres. While the technical aspects of paediatric cardiac surgery as a discipline are beyond the scope of this paper, the applied approach of systems thinking form the foundation of this reflection. This discussion will focus firstly on how previous scientific inquiry forms the basis for our current understanding in paediatric cardiac surgery. The lens of history will describe how discovery and innovation has influenced practice. To highlight the progress in our understanding as a medical community, two paediatric cardiology milestones will be discussed; specifically, the post-mortem description of an atrial septal defect (ASD) and the development of the Blalock-Taussig (BT) shunt in response to Tetralogy of Fallot. Secondly, the ‘present-day’ context will be considered. My elective at the Hospital for Sick Children provided a framework to observe and consider current technologies at our disposal to improve quality of life. Hypoplastic left heart syndrome, while not a ‘modern’ pathology, still presents a challenge to surgical practice and will be discussed. Furthermore, the left-ventricular assistance device (LVAD) will be noted as a recent innovation.

Finally, the universal demand of high-level communication will be emphasised. The need for a systematic engrained approach to multi-disciplinary team meetings, the ability to prioritise function over technical feasibility, and the aim to ensure a global standard to paediatric cardiac surgery will be discussed. Evidently, this piece has the luxury of painting with broad brushstrokes, elaborating only on a few key cases in limited detail. The purpose is to offer insight and feed the insatiable curiosity that the medical student elective experience affords.



## Body Text

### The Lens of History

Our understanding of the biological human system and our capacity to modify the human heart will always be evolving, building upon the observations of our predecessors. A historical understanding of anatomy informs us that form influences function - “simplicity is the ultimate form of sophistication” [1]. While, as a medical student in the twenty-first century, I can conceive of the anatomy involved in an atrial septal defect, this was not always the case. During the period between 1485 and 1515, Leonardo Da Vinci described in his post-mortem work *Quarderni d’Anatomica* how he had “found from … left auricle to … right auricle, a perforating channel” and questioned “whether this occurs in other auricles of other hearts” [2]. Through repeated observations, borne on the backs of many individuals over time, evidence was formed and knowledge established. With knowledge and the ability to predict and pre-empt, one paves the way for therapeutic intervention.

Emblematic of historical scientific pursuit revolutionising clinical practice is the Blalock-Taussig or “BT” shunt, a key development in the field of paediatric cardiac surgery [2]. The revolutionary paper was published in 1954, entitled “The surgical treatment of malformations of the heart in which there is pulmonary stenosis or pulmonary atresia” [3]. At the time, the challenge and clinical manifestation was known as the “blue baby,” the term conveying the sense of a fate beyond medical and surgical aid [2]. The cyanosis was observed by paediatrician Dr Helen Taussig, who collaborated with Dr Alfred Blalock and Vivien Thomas (originally a carpenter and the grandson of a slave in Louisana). This was an unusual collaboration at a time when women were rarely considered faculty and there were racially segregated wards at John Hopkins [3,4]. From the bedside to the laboratory (where the disease was modelled on animals) to the final development of surgical techniques, their systematic approach demonstrates the evolution of knowledge for practical application [3]. Today, we know that Tetralogy of Fallot is one cause of a blue baby. In Tetralogy of Fallot, blood flow to the lungs is reduced by a stenosed or atretic pulmonary valve [3]. The BT shunt provides a critical route for blood from the heart to bypass the lesion and effectively reach the lungs for oxygenation to occur [3,5]. The infants “were turned from blue to pink” [2]. This advancement won Blalock and Taussig the Nobel Prize, while Thomas, who helped design the instruments and anastomosis, received no formal recognition at the time, a reminder that the greater social context is an overarching system at play [3,6]. Today, social values have changed and the work by all three contributors lives on in a modified BT shunt used to treat congenital lesions.



**Present-Day Technology and Capability**

As we move our discussion to the present-day, the same treadmill system of knowledge rearing innovation applies. Within the last few decades, an area of development and surgical advancement is in procedures performed on single-ventricle patients [7]. Previously, children who presented with underdeveloped ‘hypoplastic’ heart chambers did not have a chance at ex utero life [7]. The disease pathogenesis is based on the embryological need of blood flow for developing cells to grow; for instance, severe congenital aortic stenosis may be the catalyst for a case of a hypoplastic left heart [8]. This affords the opportunity for minimally invasive ultrasound-guided intervention to alter the natural history and mitigate morbidity and mortality [8]. Nevertheless, there are risks associated with fetal intervention requiring both short and long-term consideration in selected patient groups [8]. Risk stratification is required in order to determine which patients will receive the optimal benefit from this evolving technology. The system of care can provide biventricular circulation and reassurance to families that their child will live through to adulthood [7]. However, single ventricle physiology persists in the population as not all cases are diagnosed prenatally, a consequence of the system of care and the tools we employ not always identifying the anatomy of the heart [7]. Given the rarity of the condition, it would not be economically feasible in our social context to screen each patient without risk stratification. Unfortunately for the treating team and families of children born with hypoplastic hearts, the post-natal surgical strategy (while offering some time and functionality) is a palliative pathway [7]. We must remember the simplicity of the heart as a service to other organ systems; biologically, a single-ventricle physiology with a decrease in cardiac output cannot sustain adequate growth and function [7].



Despite poor outcomes in some patients with congenital heart defects, decompensation does not preclude hope what with modern day technology improving outcomes. There are ongoing efforts and advances in surgical strategy to optimize the system of the heart, the body, and the patient. Indeed, previously palliated patients with progressive ventricular failure and hypoxia have been put on mechanical assistance devices as an alternative therapy [9]. The left ventricular assistance device (LVAD), also known as the “Berlin heart,” is a means of circulatory support for the failing native heart [9]. As a paracorporeal system, it is cannulated into the internal heart but rests external to the body [9]. It functions like a bike pump with a pneumatic driver and a movable membrane. Suction allows filling of the device during diastole and positive pressure facilitates ejection into the systemic circulation during systole [10]. The ingenious invention provides destination therapy as a reliable bridge to transplant or definitive treatment [10,11]. However, it is not successful in all cases. Recent multicenter data has shown the rate of survival to heart transplant or recovery with use of the LVAD to be 75% [11]. Moreover, despite reports of successful bridging to transplantation, there is limited use of ventricular assistance devices in single-ventricular patients with higher rates of mortality compared to patients with biventricular physiology [11]. Furthermore, there are complications including embolic stroke, which requires anticoagulation in all settings [9]. Another consideration is that immunological consequences of ventricular assistance devices may limit the capacity for later organ transplantation due to an increased incidence of rejection and graft loss following transplant [9]. Regardless, the LVAD has improved standard best practice care. Prior to the early 2000s, the predominant mode of circulatory support was extracorporeal membrane oxygenation which required intensive care provision [10,11]. The advent of the LVAD, which does not require intensive care, provides a more reliable alternative with improved survival, patient function, and satisfaction [10,11]. The noteworthy difference is the capacity for ambulatory care, affording the potential for an improved quality of life - the ability to mobilise, go to the ward, and play [10,11]. Moreover, there is also some consideration of a safe transition to community outpatient care [12]. In the adult population, patients on LVADs are discharged safely from hospital, however in the paediatric population there remains some trepidation because primary caregivers and schoolteachers need to be educated and adequately supported to manage risk in a more vulnerable patient group [12]. The LVAD is a testament to how we can optimise the mechanical system to support the psychosocial system within which a patient functions.



**The need for high-level communication in high-level care**

The treating team requires high-level communication because, despite technological advances, survival does not always correspond to quality of life. While the majority of babies with congenital heart disease survive, many have impaired neurodevelopment [13]. For a hospital that has a plethora of resources and expertise, knowing when to withdraw treatment is a difficult choice. As a clinical case, a patient on extra-corporeal membrane oxygenation (ECMO) did not continue to undergo active treatment, despite the technically feasible option of having an LVAD. When CT brain findings showing multiple cortical infarcts were discussed at a multi-disciplinary team [MDT] meeting, palliation was considered to be in the best interest of the patient and their family. Raising the concern highlighted the culture of the unit, which promoted open communication to balance and contrast the different system outcomes: biological, technical, and functional.

The team cohesion highlighted was fostered by the hospital system's implemented strategy. As a commitment to the ongoing improvement of the unit, weekly performance rounds made the point of retrospective analysis for each patient's clinical course. Behind the impetus to go through every patient was the recognition that these were high-risk cases with inherent potential for error and identifiable threats to patient outcomes [14]. This process of identifying and managing error and risk through effective utilization of all resources available is central to a concept known as "crew resource management" (CRM) [14]. CRM, conceived by NASA and aviation experts, is an integral part of systems thinking and is practically applied and mandated in commercial cockpit training [14]. In the hospital context, each post-operative course was considered a patient "flight," with their preoperative management strategy or "flight-plan" juxtaposed and examined with the actual clinical trajectory (plotted from the arrival in the operating room until point of discharge or death) [14]. The strategy acknowledges and learns from other high-stake industries that high-level care transcends the profession, requiring inter-disciplinary dialogue and conversation. While there is a systemic vigilance engrained, the NASA approach promotes blame-free error assessment [14]. This highlights how high-level care rests not only on technical pillars, but also on non-technical skills and system policy.



More broadly, the communication between treating centres also contributes to the promotion of high-level care. In 2017, a world database for paediatric and congenital heart surgery was established [15]. Institutions from Japan to Mexico and Colombia to Italy are communicating their practice details including demographics, pre-operative patient history, surgical data, and 1-year post-operative outcomes [15]. By confidentially comparing centre-specific data to regional, national, and international aggregated data, quality improvement strategies can be identified and implemented with guidance from international experts in the field [15]. Aligned with our societal value of universal health care, the aim is to improve outcomes on a global front [15]. This encompassing system of a worldwide standard, regardless of societal or economic status, forms the concluding sentiment of this discussion. While this paper has considered many variables in high-level paediatric care, the constant will always be the universal applicability of the human heart which can be found in any hospital, nation, or human time period.

## Conclusion

This paper has discussed many interconnected systems, telescoping through the perspectives of history, technology, and communication strategies. Ultimately, high-level care in the context of paediatric heart surgery requires a firm grasp of each system. Whether it be risk-stratification, technological innovation, or the interpretation of a worldwide surgical database, an array of connected interventions are required to optimize patient outcomes.


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