


Perfusion Management in Shone's Complex Cardiac Surgery: A Case Report

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Abstract

This case report details the advanced perfusion management of a patient presenting with Shone's Complex, a rare congenital cardiac anomaly. To minimize intraoperative hemodilution and ensure optimal organ and neuroprotection, a customized dual-oxygenator strategy was developed and successfully utilized. Systemic perfusion was maintained using a Nipro Brizio adult membrane oxygenator, while selective cerebral perfusion was independently regulated using a dedicated LILIPUT 902 pediatric oxygenator circuit. Retrograde Autologous Priming (RAP) was performed prior to the initiation of cardiopulmonary bypass to optimize intraoperative hematocrit levels. The separate circuits allowed for precise regional temperature management, maintaining a cooler cerebral environment while preserving systemic organ perfusion at a higher temperature. The patient was successfully weaned from bypass with stable hemodynamics and a postoperative hemoglobin level of 9.1 g/dL without requiring blood transfusions. Significantly, this highly sophisticated hybrid circuit configuration was established for the first time in the medical history of Afghanistan. Managed by a dedicated perfusionist adhering strictly to international standards, this milestone demonstrates that advanced, specialized perfusion frameworks can be safely executed to improve clinical outcomes in complex congenital cardiac surgeries within developing nations.

Keywords: Shone's Complex; Perfusion Management; Cardiopulmonary Bypass; Retrograde Autologous Priming (RAP); Selective Cerebral Perfusion; Neuroprotection

Introduction

Shone's Complex, alternatively known as Shone's syndrome, is an exceedingly rare and structurally devastating congenital cardiac anomaly first formally characterized by Shone and colleagues in 1963. The syndrome is classically defined by the co-existence of four obstructive left-sided lesions: a Supravalvular Mitral Ring (SVMR), a parachute-like deformity of the mitral valve, subaortic stenosis (which can present as either a discrete membranous or a muscular narrowing) and coarctation of the aorta. Managing patients with this complex constellation of anomalies into adolescence or adulthood is highly uncommon, as the vast majority of these individuals experience severe cardiac decompensation and require definitive surgical intervention during early infancy. When a patient survives to late adolescence without complete corrective surgery, the chronic volume and pressure overloads induce severe secondary structural changes. These include left ventricular hypertrophy, pulmonary hypertension and altered systemic arterial compliance, all of which present profound, multifaceted challenges to the entire clinical team [1,2].

The primary physiological and mechanical hurdles during corrective open-heart surgery for Shone's Complex revolve around maintaining stable systemic hemodynamics, ensuring optimal myocardial preservation and safeguarding the central nervous system. Cardiopulmonary Bypass (CPB) in these scenarios demands unconventional cannulation strategies, adaptive fluid dynamics and strict protocols designed to minimize blood-component trauma. Traditional, uniform single-pump CPB circuits are generally inadequate for such cases, as they fail to account for the dramatic regional pressure variations inherent to multi-level left-sided heart obstructions.

Compounding these physiological challenges, conducting such advanced maneuvers in developing healthcare infrastructures like Afghanistan presents unprecedented systemic difficulties. Decades of conflict have severely restricted the development of specialized medical fields, leaving a critical shortage of certified, highly qualified perfusionists capable of managing complex neonatal and adolescent congenital bypass setups. To bridge this gap, this case introduces a groundbreaking milestone: the implementation of an advanced hybrid perfusion circuit designed and executed by Jawad Azizi, practicing strictly in accordance with modern international perfusion standards. This report outlines the technical blueprint of this historic operation, establishing a new benchmark for cardiothoracic care in the region [3].

Case Presentation

A 16-year-old female weighing 43 kg with a known history of congenital cardiovascular anomalies presented to our institution complaining of progressive exertional dyspnea, fatigue and severely reduced exercise tolerance over the preceding six months. On clinical evaluation, her physical examination revealed a prominent systolic murmur loudest at the left sternal border and an asymmetric blood pressure gradient between her upper and lower extremities. A comprehensive diagnostic workup, including transthoracic echocardiography and Computed Tomography (CT) angiography, was performed. The imaging confirmed classic features consistent with Shone's Complex, demonstrating severe left-sided obstructive cardiac lesions, an abnormal mitral valve subvalvular apparatus with a single dominant papillary muscle cluster (indicative of a parachute mitral valve), a distinct subaortic fibrous ring causing a high left ventricular outflow tract gradient and an upper aortic arch coarctation [1,2].

Given the severity of the multi-level obstructions and the high risk of impending heart failure, the patient was scheduled for urgent, comprehensive surgical correction under cardiopulmonary bypass. The procedure was scheduled at the Ariana Medical Complex in Kabul, Afghanistan. Preoperative laboratory investigations revealed a stable baseline hematocrit and renal function, but given her relatively low body weight and small circulating blood volume, the clinical team identified a high risk for severe intraoperative hemodilution if standard adult bypass techniques were utilized. A multidisciplinary clinical planning session involving cardiovascular surgeons, cardiac anesthesiologists and the perfusion team concluded that a standard, off-the-shelf single-pump bypass circuit would not provide adequate safety margins for the regional flow demands and neuroprotective parameters required for this complex repair.

Perfusion Management

Perfusion management was meticulously designed and conducted by Jawad Azizi using a customized hybrid circuit designed to optimize blood conservation and cerebral protection.

A. Dual-Circuit Configuration

Two separate, independent membrane oxygenator circuits were integrated into a single hybrid system to meet the divergent physiological requirements of the patient's anatomy. A pediatric circuit was dedicated exclusively to selective cerebral perfusion, allowing the team to maintain precise, low-flow, low-pressure delivery to the carotid vessels during periods of systemic adjustment. Simultaneously, systemic metabolic demand and abdominal organ perfusion were continuously maintained using a high-efficiency Nipro Brizio adult membrane oxygenator. This mechanical separation prevented pressure spikes within the cerebral vasculature while ensuring that the rest of the body received optimal caloric and oxygen delivery throughout the extensive cross-clamp period.

B. Reservoir Optimization

To minimize the circuit's total priming volume and directly limit the severity of intraoperative hemodilution, a LILIPUT 902 pediatric reservoir was strategically incorporated into the systemic circuit layout instead of using a standard, high-volume adult venous reservoir. This modification reduced the baseline static volume required to safely operate the circuit, effectively lowering

the amount of crystalloid fluid that would mix with the patient's native circulating blood volume upon the initiation of cardiopulmonary bypass.

C. *Selective Cerebral Perfusion Strategy*

A dedicated, secondary roller pump head enabled independent control of cerebral flow rates, driving pressures and perfusate temperature. This mechanical layout facilitated a precise "cold head–warm body" perfusion approach. The cerebral circuit was cooled to achieve profound localized hypothermia, maximizing neurological cellular protection and reducing cerebral metabolic oxygen demand. Meanwhile, the systemic circuit was maintained at a significantly milder hypothermic level, protecting systemic abdominal organs from ischemic stress while avoiding the profound systemic coagulopathies, prolonged rewarming times and myocardial stunning typically associated with deep hypothermia.

D. *Blood Conservation (RAP Technique)*

To protect the patient's native red blood cell concentration, Retrograde Autologous Priming (RAP) was rigorously performed prior to going on bypass. This technique involved carefully utilizing the patient's own arterial and venous pressures to retrogradely displace approximately 500 mL of the clear crystalloid priming fluid out of the circuit lines and into a sterile collection bag. This displaced volume was replaced entirely by the patient's native blood before the main pumps were activated. Consequently, this maneuver effectively prevented the severe drop in hematocrit that routinely occurs when a low-weight patient is connected to an unprimed or crystalloid-primed extracorporeal circuit.

Surgical Procedure

Following the induction of general anesthesia and routine monitoring (including invasive arterial lines in both the right radial artery and femoral artery to track regional pressures), a standard median sternotomy was performed. The pericardium was opened and suspended to expose the severely distorted cardiac anatomy. High ascending aortic cannulation was carefully executed alongside dual stage cavatrial cannulation to establish a robust venous return line. Once the specialized hybrid cardiopulmonary bypass circuit was fully integrated, the patient was progressively cooled toward a target core systemic temperature.

The surgical repair demanded a meticulous, staged approach to address the multiple left-sided obstructive pathologies. After applying the aortic cross-clamp, cold blood cardioplegia was delivered directly into the aortic root to achieve complete cardiac arrest and optimize myocardial protection. The procedure was performed by Dr. Khaiber Sidiqi, with anesthesia managed comprehensively by Dr. Shabir Azizi. Surgical correction addressed the left-sided lesions under cardiopulmonary bypass support. The surgeon first addressed the subaortic area, carefully resecting the obstructive subaortic membrane to widely clear the Left Ventricular Outflow Tract (LVOT). Attention was then turned to the mitral apparatus via a left atriotomy. The supra-annular mitral ring was precisely excised and the chordae of the parachute mitral valve were carefully inspected and fenestrated to relieve the subvalvular inflow gradient without inducing severe insufficiency. Perfusion stability was tightly regulated throughout these delicate maneuvers using a localized, independent pump architecture to prevent systemic structural collapse [3].

Outcome

Following the successful anatomical resection of the obstructive membranes and the repair of the valvular structures, the patient was progressively and safely rewarmed. Myocardial contractility returned vigorously following the removal of the cross-clamp. The patient was successfully weaned from cardiopulmonary bypass without any hemodynamic instability or rhythm complications. Hemodynamic parameters remained perfectly stable throughout the immediate post-bypass phase and the aggressive blood conservation protocol yielded an excellent postoperative hemoglobin level of 9.1 g/dL on the operating table. The patient was transferred to the intensive care unit in stable condition, extubated early the following morning and experienced an entirely uneventful recovery with a satisfactory clinical outcome. Follow-up echocardiography confirmed the complete elimination of the subaortic gradient and a significantly improved flow profile across the mitral valve.

Discussion

The successful surgical correction of a 16-year-old presenting with advanced Shone's Complex highlights the vital necessity of individualized, case-specific perfusion algorithms over standard institutional configurations. The structural anomalies of Shone's

syndrome create an environment where traditional single-pump systems present high risks of regional hypoperfusion or cerebral hyperbaric trauma. By utilizing a customized dual-circuit configuration—separating an adult Brizio membrane oxygenator for systemic metabolic demands from a pediatric Liliput 902 circuit dedicated exclusively to selective cerebral perfusion—we successfully decoupled systemic metabolic support from delicate cerebral autoregulation mechanics. This configuration permitted a "cold head-warm body" strategy, providing critical neuroprotection to the brain via deep hypothermia while avoiding the systemic metabolic derangements, coagulopathies and prolonged rewarming phases associated with profound whole-body hypothermia [4].

Furthermore, fluid shift mitigation and blood conservation remain paramount when operating on low-weight adolescents undergoing complex open-heart surgery. Implementing Retrograde Autologous Priming (RAP) allowed for the safe displacement of approximately 500 mL of crystalloid prime using the patient's own native blood prior to bypass. This maneuver preserved baseline red blood cell mass, stabilized intraoperative oncotic pressure and yielded an excellent postoperative hemoglobin level of 9.1 g/dL without requiring external donor red cells.

The broader clinical significance of this case extends far beyond its physiological successes, representing a historic triumph for the medical infrastructure of Afghanistan. Decades of geopolitical instability, war and structural isolation have profoundly crippled the nation's healthcare system. Consequently, the country has severely lacked qualified perfusion professionals possessing the advanced training required to construct non-traditional, multi-pump circuits. Most local surgeries have historically been limited to basic, single-circuit adult configurations due to a lack of specialized training and resource constraints. This operation marks the first time in Afghan medical history that a hybrid, dual-oxygenator perfusion circuit was successfully designed, primed and operated. By strictly implementing international clinical guidelines, utilizing precise safety checklists and adapting available pediatric and adult technologies, Jawad Azizi demonstrated that world-class, standard-compliant perfusion safety can be achieved even within a deeply recovering healthcare ecosystem. This historic achievement serves as a reproducible model for other cardiothoracic teams operating in resource-limited or conflict-affected regions globally [5-7].

Conclusion

Precision perfusion management, including the utilization of Retrograde Autologous Priming (RAP) and targeted neuroprotective dual-circuit strategies, is essential when managing complex congenital cardiac anomalies like Shone's Complex. Individualized perfusion planning significantly improves intraoperative safety margins, protects vital end-organs and minimizes the need for allogeneic blood products. This historic case marks the first time a specialized hybrid circuit setup was successfully executed in Afghanistan, proving that adherence to rigid international standards by dedicated clinical professionals can overcome deep structural and post-war healthcare limitations to deliver elite patient care.

Conflict of Interest

The authors declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

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The data supporting the findings of this study are available from the corresponding author upon reasonable request.

Ethical Statement

The project did not meet the definition of human subject research under the purview of the IRB according to federal regulations and therefore was exempt.

Informed Consent Statement

Informed consent was obtained from all participants included in the study.

Authors' Contributions

All authors contributed equally to this paper.

References

1. Dearani JA, Danielson GK, Puga FJ, Schaff HV, Warnes CA. Surgical management of Shone's complex and related congenital left-sided obstructive lesions. *Ann Thorac Surg.* 1999;68:1357-63.
2. Gravlee GP, Davis RF, Kurusz M, Utey JR. *Cardiopulmonary bypass: Principles and practice.* 3rd Ed. Philadelphia: Lippincott Williams and Wilkins. 2008.
3. Jonas RA. *Comprehensive surgical management of congenital heart disease.* 2nd Ed. London: Hodder Arnold. 2014.
4. Kurusz M, Butler BD, Katz J, Conti VR. Techniques for minimizing hemodilution during cardiopulmonary bypass. *Perfusion.* 2000;15:187-92.
5. Groom RC, Forest RJ. Retrograde autologous priming for cardiopulmonary bypass: Techniques and benefits. *J Extra Corpor Technol.* 2001;33:38-42.
6. Bical OM, Fromes Y, Gaillard D, Fischer M. Selective cerebral perfusion and neuroprotection in complex congenital cardiac surgery. *Eur J Cardiothorac Surg.* 2002;21:538-43.
7. Hraska V. Perfusion strategies in pediatric and congenital cardiac surgery. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu.* 2013;16:45-50.

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