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Case Report

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Spinal Anesthesia for Urologic Surgery in an Infant with Hemi-Fontan Physiology: A Case Report

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Abstract
Hypoplastic left heart syndrome is a congenital heart defect characterized by hypoplasia of left-sided heart structures and a single functional right ventricle. The resultant physiology poses significant challenges for the anesthesiologist when general anesthesia is necessary for these children. This case report describes an infant with hemi-Fontan physiology undergoing circumcision in which awake spinal anesthesia was used with favorable hemodynamic, respiratory, and surgical conditions. Written consent for publication was obtained from the patient's parents.

Keywords: Congenital Heart Disease, Neurodevelopment, Local Anesthetics, Infant

Introduction
Hypoplastic left heart syndrome (HLHS) is characterized by complete mixing of arterial and venous blood and distribution to systemic and pulmonary circulations by the right ventricle [1]. Surgical repair typically occurs in three stages: Norwood, hemi-Fontan or bidirectional Glenn, and Fontan procedures. This case report describes an infant with hemi-Fontan physiology presenting for circumcision performed safely under spinal anesthesia (SA).

Case Description
A 16-month old, 10.4 kg, 83 cm full-term male with HLHS presented for circumcision. Past medical history included HLHS, status-post Norwood and hemi-Fontan procedures, with normal coagulation status. He displayed moderate functional capacity with baseline oxygen saturation of 85%-89%. Pre-operatively, 4% lidocaine cream was applied to the patient's skin at L3-5 and 7.8 mg of oral midazolam was administered. In the operating room, standard monitors were placed.

1 mL of hyperbaric 0.5% tetracaine with clonidine 10 mcg was injected via a 1.5-inch Sprotte needle at the L3/4 interspace. Next, an intravenous catheter was placed in the patient's foot for administration of maintenance fluids and rescue medications if needed. Sensory clamp test was negative, and the patient fell asleep prior to incision. Hemodynamics remained within 20% of the patient's baseline during surgery (see Figure 1). Normal lower extremity strength returned 95 minutes after spinal injection. The patient was discharged home with surgical penile block and oral acetaminophen 160 mg for post-operative pain control.

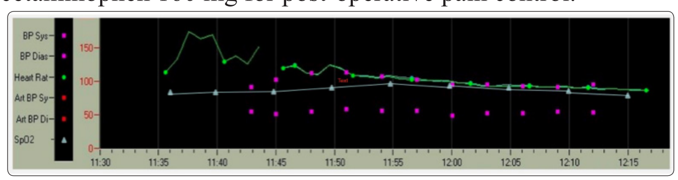

Figure 1: Intraoperative vital signs during placement of spinal and throughout surgery.

Discussion
Anesthesia for non-cardiac surgery in patients with HLHS
Neonates with HLHS typically undergo a Norwood procedure, which consists of neoaorta creation, atrial septectomy, and creation of a shunt to provide pulmonary blood flow (PBF). Systemic blood flow (SBF) and PBF become parallel circuits supported by the single ventricle; thus, patients with Norwood physiology are sensitive to alterations in pulmonary vascular resistance (PVR), preload, and afterload and are at highest risk of perioperative complications [1].

Around six months of age, the superior vena cava (SVC) is connected to a branch pulmonary artery in the hemi-Fontan or bidirectional Glen procedure, establishing a dedicated source of PBF and offloading the single ventricle. Thereafter, gradient-driven PBF requires maintenance of adequate preload and avoidance of significant increases in PVR [1].

Finally, around two years of age, the inferior vena cava (IVC) is redirected to the pulmonary circulation in the Fontan procedure.

After this stage, patients are sensitive to decreases in cardiac output; thus, adequate preload, contractility, and sinus rhythm must be maintained [1].

Children with CHD requiring anesthesia for non-cardiac surgery have a two-fold higher rate of cardiac arrest and death during the perioperative period; patients with single ventricle physiology are among those with the highest risk [2]. Of the three stages, bidirectional Glenn or hemi-Fontan physiology is considered to be the most hemodynamically-stable, resulting from reduced volume load on the single ventricle compared with Norwood physiology as well as maintenance of preload from the IVC compared with Fontan physiology [1].

Spinal anesthesia for patients with congenital heart disease

Use of SA in infants and toddlers is associated with cardiovascular and respiratory stability [3]. Reduced baseline sympathetic activity with a balanced reflex reduction in vagal activity may explain the lack hemodynamic perturbations seen in young children [3]. Furthermore, several studies have shown that this hemodynamic stability is preserved specifically in patients with CHD; thus the hemodynamic stability afforded by SA makes it a desirable approach for single ventricle patients [2].

Avoiding tracheal intubation and preserving spontaneous ventilation may also improve overall perioperative stability in HLHS patients. As PBF is gradient-driven in the Hemi-fontan or bidirectional Glenn patient, avoiding positive pressure ventilation and increases in intrathoracic pressure is preferred. Additionally, awake SA is associated with a decreased rate of apnea compared to general anesthesia (GA) in infants without a history of apnea and who do not receive peri-operative sedatives [4].

SA also avoids the use of potentially neurotoxic general anesthetic agents. While several recent studies have concluded that one anesthetic of short duration is likely safe for children, risk of neurotoxicity after multiple, prolonged anesthetic exposures is less clear [5]. This is an important consideration in patients with HLHS who require several, prolonged anesthetics early in life. Avoidance of GA, when possible, may be of additional benefit in this population.

In summary, young children with HLHS are at high risk of cardiac and respiratory complications related to GA. SA, with its associated hemodynamic stability, preservation of spontaneous ventilation, and avoidance of potentially neurotoxic agents, may be a preferable anesthetic technique for this unique patient population.

Learning Points

1. Patients with congenital heart disease, especially those with HLHS, are at increased risk of adverse events related to general anesthesia for non-cardiac surgeries.
2. Spinal anesthesia preserves spontaneous ventilation and hemodynamic stability in patients with HLHS.
3. Use of spinal anesthesia for patients with congenital heart disease avoids exposure to volatile anesthetics and other potentially neurotoxic agents.

Ethics: Parental permission obtained.

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