



ABSTRACT

Hypoplastic Left Heart Syndrome (HLHS) is a congenital heart defect whereby the left-sided structures of the heart are underdeveloped, leading to compromised systemic blood flow. Children with HLHS often require placement under general anesthesia to complete treatment; however, care must be taken in order to ensure safe outcomes, as these children are at an increased risk for developing perioperative complications. The purpose of this case report is to discuss the medical management of a child with HLHS requiring dental treatment under general anesthesia. This case demonstrates that multidisciplinary collaboration attained a positive outcome even when faced with sustained perioperative hypotension.

INTRODUCTION

Hypoplastic Left Heart Syndrome (HLHS) is a congenital heart defect characterized by the underdevelopment of the left-sided structures of the heart, including the mitral valve, left ventricle, aortic valve, ascending aorta, and aortic arch.^{1,4,5} Making up about 7-9% of all congenital heart disease, males have been demonstrated to have a greater predilection to develop this condition over females (1.5:1).^{1,3} More importantly, HLHS is responsible for 23% of cardiac deaths during the first week of life, making this condition the leading cause of neonatal mortality.¹

Given that these children suffer from either an absent or insufficiently-sized left ventricle, this defect leads to a significant deficit in oxygenated blood being pumped into the systemic circulation. Their smaller-sized aorta also places them at high risk for developing aortic valve atresia with atresia/severe stenosis of the mitral valve, leading to overall poor systemic blood perfusion.^{1,4,5} Atrial septal defects are also common in these children as they serve as nature's way of compensating for the backlog of oxygenated blood. Due to the mixing of oxygenated and deoxygenated blood, systemic blood saturation is typically diminished to 75-85% within these children.

A multidisciplinary team approach is essential for the successful management of HLHS. The current management strategy of children afflicted with HLHS includes: a lifetime follow-up with a cardiologist, a multi-targeted cardiac medication regimen, and surgical intervention when indicated.

As these children bear such a significant cardiac burden, their medical procedures are typically completed under general anesthesia as a means at promoting the safe delivery of their care. However, careful consideration is warranted as these children pose an increased risk for developing complications like cardiogenic shock, cyanosis and arrhythmias during surgery.¹ Therefore, whether for dental or medical treatment under general anesthesia, these children require cross-functional team coordination at minimizing such complications from occurring.

This case report documents the perioperative management of severe hypotension in a child with HLHS requiring dental treatment under general anesthesia

CASE REPORT

A 9-year 9-month old female was referred for restorative treatment under general anesthesia. Her medical history consists of: DiGeorge syndrome, HLHS, G-Tube dependence, delayed bone age, and protein losing enteropathy. She takes aspirin, budesonide, enalapril, and spironolactone daily for management of HLHS. She has no known drug allergies.

She has undergone multiple cardiac surgeries to correct her congenital heart defect. Given that she has had multiple exposures to general anesthesia with successful outcomes, the possibility for an adverse event to occur appeared relatively unlikely. However, the relative risk still remained.

Despite her short stature, her general appearance was unremarkable. No pathology was detected either extra- or intra-orally at day of surgery. Overall, she had fair oral hygiene. She presented in the mixed dentition stage and her dental development appeared to be consistent with her chronological age. She had three over-retained primary teeth. She presented with bilateral class 1 molar and canine occlusion, with an overbite of 15% and an overjet of 3 mm (Figure 1).

DIAGNOSTIC ASSESSMENT

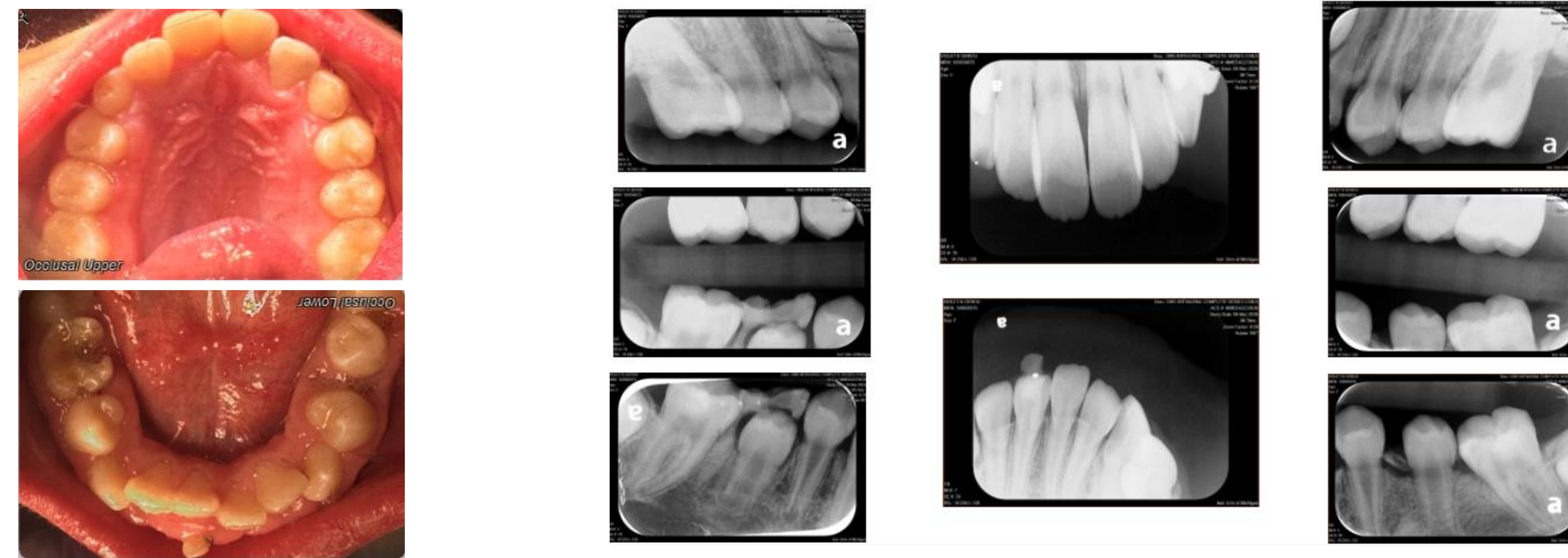


Figure 1: Intraoral Photographs & Full Mouth Series

Standard	155 -DU	153 -DU	102 -DU	96 -DU	95 -DU
HR-EKG	155 -DU	153 -DU	102 -DU	96 -DU	95 -DU
Row Name	03/09/26 0822	03/09/26 0821	03/09/26 0820	03/09/26 0819	03/09/26 0818
Agents					
O2 Flow	0.37 L/min -DU	0.37 L/min -DU	0.37 L/min -DU	0.37 L/min -DU	0.37 L/min -DU
Air Flow	0.63 L/min -DU	0.63 L/min -DU	0.63 L/min -DU	0.63 L/min -DU	0.63 L/min -DU
N2O Flow	0 L/min -DU	0 L/min -DU	0 L/min -DU	0 L/min -DU	0 L/min -DU
Nitrous Insp	0.1 % -DU	0.1 % -DU	0.1 % -DU	0.1 % -DU	0.1 % -DU
Nitrous Exp	0 % -DU	0 % -DU	0 % -DU	0 % -DU	0 % -DU
Sevo Insp	1.5 % -DU	1.8 % -DU	1.8 % -DU	1.9 % -DU	1.9 % -DU
Sevo Exp	1.4 % -DU	1.6 % -DU	1.6 % -DU	1.6 % -DU	1.8 % -DU
Temp					
Rectal Temp	35.7 °C (96.3 °F) -DU	35.7 °C (96.3 °F) -DU	35.7 °C (96.3 °F) -DU	35.7 °C (96.3 °F) -DU	35.7 °C (96.3 °F) -DU
Cardiac					
HR-SpO2	95 -DU	98 -DU	109 -DU	96 -DU	98 -DU
NIBP	60/37 † -DU	41/15 † -DU	—	54/21 † -DU	—
NIBP Mean	—	43 -DU	22 -DU	—	30 -DU
Resp					
SpO2	95 % -DU	94 % -DU	87 % † -DU	93 % -DU	92 % -DU
End Tidal CO2	41.8 mmHg -DU	42.6 mmHg -DU	44 mmHg -DU	46.2 mmHg -DU	36.7 mmHg -DU

Standard	155 -DU	155 -DU	155 -DU	153 -DU	154 -DU
HR-EKG	155 -DU	155 -DU	155 -DU	153 -DU	154 -DU
Row Name	03/09/26 0827	03/09/26 0826	03/09/26 0825	03/09/26 0824	03/09/26 0823
Agents					
O2 Flow	0.37 L/min -DU	0.37 L/min -DU	0.37 L/min -DU	0.37 L/min -DU	0.37 L/min -DU
Air Flow	0.63 L/min -DU	0.63 L/min -DU	0.63 L/min -DU	0.63 L/min -DU	0.63 L/min -DU
N2O Flow	0 L/min -DU	0 L/min -DU	0 L/min -DU	0 L/min -DU	0 L/min -DU
Nitrous Insp	0 % -DU	0 % -DU	0 % -DU	0 % -DU	0.1 % -DU
Nitrous Exp	0 % -DU	0 % -DU	0 % -DU	0 % -DU	0 % -DU
Sevo Insp	1.2 % -DU	1.2 % -DU	1.3 % -DU	1.3 % -DU	1.4 % -DU
Sevo Exp	1.1 % -DU	1.2 % -DU	1.2 % -DU	1.3 % -DU	1.3 % -DU
Temp					
Rectal Temp	35.8 °C (96.4 °F) -DU	35.8 °C (96.4 °F) -DU	35.8 °C (96.4 °F) -DU	35.7 °C (96.3 °F) -DU	35.7 °C (96.3 °F) -DU
Cardiac					
HR-SpO2	157 -DU	154 -DU	104 -DU	98 -DU	96 -DU
NIBP	—	77/40 † -DU	53/25 † -DU	—	50/19 † -DU
NIBP Mean	—	54 -DU	33 -DU	—	28 -DU
Resp					
SpO2	95 % -DU	95 % -DU	94 % -DU	95 % -DU	96 % -DU
End Tidal CO2	44.7 mmHg -DU	44.8 mmHg -DU	43.3 mmHg -DU	41.1 mmHg -DU	41.8 mmHg -DU

Standard	150 -DU	156 -DU	168 -DU	162 -DU	155 -DU
HR-EKG	150 -DU	156 -DU	168 -DU	162 -DU	155 -DU
Row Name	03/09/26 0832	03/09/26 0831	03/09/26 0830	03/09/26 0829	03/09/26 0828
Agents					
O2 Flow	0.5 L/min -DU	0.37 L/min -DU	0.37 L/min -DU	0.37 L/min -DU	0.37 L/min -DU
Air Flow	0 L/min -DU	0.63 L/min -DU	0.63 L/min -DU	0.63 L/min -DU	0.63 L/min -DU
N2O Flow	1.5 L/min -DU	0 L/min -DU	0 L/min -DU	0 L/min -DU	0 L/min -DU
Nitrous Insp	9.7 % -DU	0 % -DU	0 % -DU	0 % -DU	0 % -DU
Nitrous Exp	1.4 % -DU	0 % -DU	0 % -DU	0 % -DU	0 % -DU
Sevo Insp	1 % -DU	1.1 % -DU	1.1 % -DU	1.1 % -DU	1.2 % -DU
Sevo Exp	0.9 % -DU	1 % -DU	1 % -DU	1 % -DU	1.1 % -DU
Temp					
Rectal Temp	35.8 °C (96.4 °F) -DU	35.8 °C (96.4 °F) -DU	35.8 °C (96.4 °F) -DU	35.8 °C (96.4 °F) -DU	35.8 °C (96.4 °F) -DU
Cardiac					
HR-SpO2	156 -DU	157 -DU	157 -DU	157 -DU	155 -DU
NIBP	56/31 † -DU	—	—	72/37 † -DU	74/42 † -DU
NIBP Mean	39 -DU	—	—	49 -DU	54 -DU
Resp					
SpO2	95 % -DU	95 % -DU	95 % -DU	95 % -DU	96 % -DU
End Tidal CO2	46.2 mmHg -DU	44 mmHg -DU	44 mmHg -DU	44.7 mmHg -DU	44.8 mmHg -DU

Figure 2: Intraoperative Vitals Demonstrating Sustained Hypotension

Standard	136 -DU	144 -DU	150 -DU	156 -DU	149 -DU
HR-EKG	136 -DU	144 -DU	150 -DU	156 -DU	149 -DU
Row Name	03/09/26 0837	03/09/26 0836	03/09/26 0835	03/09/26 0834	03/09/26 0833
Agents					
O2 Flow	0.5 L/min -DU	0.5 L/min -DU	0.25 L/min -DU	0.5 L/min -DU	0.5 L/min -DU
Air Flow	0 L/min -DU	0 L/min -DU	0 L/min -DU	0 L/min -DU	0 L/min -DU
N2O Flow	1.5 L/min -DU	1.5 L/min -DU	0.75 L/min -DU	1.5 L/min -DU	1.5 L/min -DU
Nitrous Insp	71.2 % -DU	70.5 % -DU	48.6 % -DU	73.7 % -DU	71.3 % -DU
Nitrous Exp	64.1 % -DU	61.8 % -DU	46 % -DU	61 % -DU	52.1 % -DU
Sevo Insp	0.3 % -DU	0.3 % -DU	0.6 % -DU	0.2 % -DU	0.2 % -DU
Sevo Exp	0.4 % -DU	0.5 % -DU	0.6 % -DU	0.4 % -DU	0.5 % -DU
Recurring Monitoring					
TOF (Manual)	—	—	—	0/4 -MH	—
EKG	—	—	—	NSR -MH	—
Eyes	—	—	—	Eyes Checked -	—
Temp					
Rectal Temp	35.9 °C (96.6 °F) -DU	35.9 °C (96.6 °F) -DU	35.9 °C (96.6 °F) -DU	35.8 °C (96.4 °F) -DU	35.8 °C (96.4 °F) -DU
Cardiac					
HR-SpO2	161 -DU	158 -DU	166 -DU	166 -DU	157 -DU
NIBP	58/26 † -DU	—	53/26 † -DU	—	—
NIBP Mean	36 -DU	—	35 -DU	—	31 -DU
Resp					
SpO2	92 % -DU	93 % -DU	95 % -DU	93 % -DU	94 % -DU
End Tidal CO2	47.6 mmHg -DU	47.7 mmHg -DU	47.7 mmHg -DU	49.8 mmHg -DU	48.4 mmHg -DU

OUTCOME(S)

A preoperative management plan was created through collaboration between cardiology, dental and anesthesia teams. The plan included: SBE prophylaxis coverage, care coordination with cardiac anesthesia, and availability of vasoconstrictors during surgery.

Her pre-procedural blood pressure was 99/46. She sustained low blood pressure values throughout dental surgery despite the use of multiple vasoconstrictors. Epinephrine was dosed over five boluses with little improvement observed. Even by doubling the dosage and rate of epinephrine, she remained significantly hypotensive (Figure 2). Vasopressin was later administered as an attempt to promote spontaneous circulation and improve her blood pressure, which was trending towards hypotensive crisis levels.

Despite all of these efforts, her blood pressure remained hypotensive, warranting admission following dental surgery. The goal for discharge was reaching a 75 systolic blood pressure. During admission, she did not require supplemental oxygen as her SpO2 levels remained adequate. However, cardiac anesthesia determined to withhold one of her cardiac medications as an attempt to optimize her blood pressure. Following 21 hours of admission, she achieved normal blood pressure levels and was discharged in satisfactory condition. A recall appointment was recommended six months after dental treatment.

DISCUSSION

Given the nature of this complex, single ventricle heart disease, afflicted children have an increased risk of developing bacteremia. Not only is SBE prophylaxis often indicated, but meticulous oral hygiene habits are of the utmost importance. It has been demonstrated that the presence of gingival bleeding after toothbrushing has been associated with almost an eightfold increase in developing bacteremia⁶. Thus, by maintaining a strong oral hygiene regimen, the risk for bacteremia can be greatly mitigated.

A multidisciplinary team approach involving cardiology, anesthesiology, and pediatric dentistry helped to manage the medical emergency of sustained hypotension during dental surgery. By following the peri-operative recommendations provided by her cardiologist, utilizing cardiac anesthesia for the delivery of her care helped to ensure a non-fatal outcome. Keeping epinephrine and vasopressin at the ready was critically important during her surgery, as these medications were used in multiple instances in an attempt to reverse her hypotensive crisis. However, despite these efforts, she required further monitoring, and ultimately required admission in order to resolve her sustained hypotension. This case report demonstrates how children with HLHS can present with compromised cardiac function that can lead to adverse events during and even after surgery. However, by combining vigilant cardiac anesthesia protocols with effective multidisciplinary collaboration, surgical risks for these children can be successfully minimized¹.

CONCLUSION

HLHS is a severe congenital heart defect where oxygenated blood cannot be effectively pumped into the systemic circulation due to the underdevelopment of left-sided structures, leading to significant cardiac compromise for these children^{1,4,5}. This case report discussed the medical management of a 9-year 9-month old female diagnosed with HLHS undergoing dental treatment under general anesthesia. With a pre-operative management plan in place, her declining blood pressure was quickly identified, enabling a nonfatal outcome. Coordinated medical care and the availability of vasoconstrictors proved to be essential entities in the management of HLHS during surgery. This report demonstrates that dental treatment can be effectively performed on children with HLHS as long as necessary precautions are taken.

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