EAS Journal of Anaesthesiology and Critical Care
Abbreviated Key Title: EAS J Anesthesiol Crit Care
ISSN: 2663-094X (Print) & ISSN: 2663-676X (Online)
Published By East African Scholars Publisher, Kenya

OPEN ACCESS

Volume-7 | Issue-4 | Jul-Aug-2025 | DOI: https://doi.org/10.36349/easjacc.2025.v07i04.003

Case Report

Anesthesia in an Child with a Single Atrium: A Case Report and Review of the Literature

Amadou HS[1*], Mahamane YS[1], Hassane ML[2], Bachar LO[1], Alfred Dogbe YZ[3], Niandou M[1], Sabo R[1], Bandiaré M[1], Foumakoye A[1], Chaibou MS[3]

[1]Amirou Boubacar Diallo National Hospital
[2]André Salifou University
[3]Niamey National Hospital

Article History
Received: 11.06.2025
Accepted: 06.08.2025
Published: 12.08.2025

Journal homepage:
https://www.easpublisher.com

Quick Response Code

Abstract: Single atrium is a rare congenital heart disease defined by a complete absence of the interventricular septum and endocardial cushion, an absence of ventricular septal defect, and an absence of atrioventricular valve malformations. Given the epidemiological, diagnostic, and therapeutic interest, and more specifically, anesthesiological management, we report a clinical case of a 9-month-old infant admitted for the treatment of stomal prolapse. The infant has several congenital malformations: a single atrium, an anorectal malformation, and hexadactyly of both thoracic and pelvic limbs. The paraclinical biological and biochemical assessments were normal. One intraoperative incident involving constant desaturation followed by cardiac arrest was reported, which progressed favorably without neurological sequelae.

Keywords: Single atrium, Congenital heart disease, Anesthesia, Anorectal malformation, Cardiorespiratory arrest.

INTRODUCTION

Single atrium is a rare congenital heart disease. It is defined by the complete absence of the interatrial septum, the absence of atrioventricular malformations, and the absence of interventricular communication. It can be isolated or associated with other heart diseases or malformations in general. It may be considered a multiple congenital malformation. Anesthesia in patients with congenital heart disease for non-cardiac surgery is uncommon. However, the lack of diagnosis and management of patients with congenital heart disease poses a challenge in our region.

CLINICAL CASE

Nine (9)-months-old child, referred for stomal prolapse, with a history of two surgical procedures on the second day of life and at nine months of age, respectively, for evacuating colostomy and anorectal plasty without prior cardiac investigation. Episodes of desaturations and recovered cardiorespiratory arrests were reported during these procedures.

These cardiorespiratory arrests recovered without neurological sequelae. On clinical examination, the infant was in good general condition, with moderately colored conjunctiva and mucous membranes, weighing 10 kg, a pulse rate of 96 pulses/mm, a pulse rate of 95% oxygen saturation on room air, no dyspnea, regular heart sounds, no murmur, and clear lungs.

Paraclinically, she is blood group O positive, blood count shows hyperleukocytosis at 18,91.10^3, anemia at 10.3 g/dl, low hematocrit at 32.9%, thrombocytosis at 635.10^3/ul, prothrombin rate at 100%, activated cephalin rate at 25s, azotemia at 0.38 g/l, creatinine at 4.06 mg/dl, fasting blood sugar at 0.83 g/dl.

As part of the malformation assessment, hexadactyly was noted in all four limbs.

Cardiac ultrasound showed "a single atrium, with good biventricular systolic function with LVEF at 62.6%, minimal mitral regurgitation, dilated right and left ventricles, an absent interatrial septum, normal venous and pulmonary return, and no PAH."

***Corresponding Author:** Amadou HS
Amirou Boubacar Diallo National Hospital

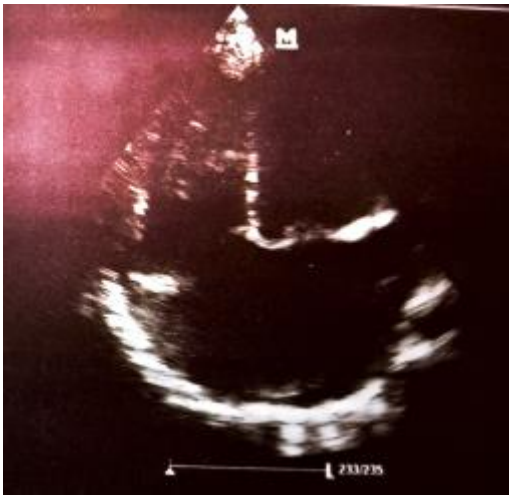

Figure 1: Single atrium visible on transthoracic echocardiography

The patient was ASA class III.

A pre-anesthetic visit was performed without any unusual findings at the patient's bedside. In the operating room, heart rate, saturation, and ECG were monitored. Vital signs at the start of the procedure included a tachycardia of 140 pulses/min, and a pulse oxygen saturation on room air of 96%.

Drugs used at induction: Fentanyl 30 µg, propofol 50 mg then 10 mg.

Four major accidents occurred during the operation: cardiac arrest during induction, rapid intubation, external cardiac massage, adrenaline, recovery, followed by constant desaturation with an SpO2 below 70% with cyanosis, then two other cardiac arrests recovered intraoperatively.

The surgery was cut short, the patient was awakened and extubated on the operating table and admitted to the post-surgical intensive care unit.

The patient was transferred to the pediatric surgery department 48 hours later, with good general condition and no neurological sequelae.

DISCUSSION

In the literature, there are two terms that can lead to confusion: single atrium and common atrium. In a single atrium, here is a complete absence of the interatrial septum and endocardial cushion, an absence of ventricular septal defect, and an absence of atrioventricular valve malformation [1].

On the other hand, a common atrium is characterized by a complete absence or the presence of a strand of tissue present at the upper atrial wall of the common chamber, an absence of ventricular septal defect, and accompanied by a defect in the atrioventricular cushion [2-4]. The pathophysiology and clinical signs are similar to those of a large atrial septal defect.

In patients with normal pulmonary artery pressure, left-to-right shunting is the most common. In patients with pulmonary hypertension, pulmonary resistance is higher than systemic vascular resistance, leading to right-to-left shunting and Eisenmenger syndrome. Exercise intolerance, palpitations and recurrent respiratory tract infections, shortness of breath, developmental delay, and other nonspecific clinical manifestations may be noted.

A soft murmur is often heard at the pulmonary site [5,6].

Eisenmenger syndrome is defined as shunt inversion with cyanosis, right heart failure, a prominent S2 burst at the pulmonary focus, and polycythemia. On radiography, the proximal pulmonary arteries are extremely dilated, with very poor peripheral pulmonary vascularization [7].

As the disease progresses, cyanosis, clubbing of the fingers and toes, and an ejection murmur in the region of the pulmonary valve may occur. In advanced disease, congestive heart failure, jugular vein distension, and liver enlargement will develop. Blood admixture at the atrial level is significantly greater in the common atrium due to atrioventricular valve regurgitation.

General anesthesia with orotracheal intubation was our choice given the surgical context and the pediatric setting. Hong C. *et al.,* had an adult subject and chose locoregional anesthesia or an epidural [1,5]. In general anesthesia, to avoid decompensation, a constant balance between systemic and pulmonary vascular resistance must be maintained. Regarding systemic vascular resistance, factors that increase it are hypoxia, hypercapnia, acidosis, sympathetic stimulation, hypervolemia, and high FiO2.

In newborns, pulmonary vascular resistance is high, and shunting is minimal. At two months of age, pulmonary vascular resistance decreases. Pulmonary

flow can reach four times the systemic flow, leading to PAH and heart failure progressing to fixed PAH, shunt reversal, or Eisenmenger complex).

The goal of anesthesia is to limit the shunt by increasing pulmonary vascular resistance (increased insufflation pressure, PEEP, moderate hypercapnia; avoid hypocapnia and hyperoxia) and by decreasing systemic vascular resistance by administering deep anesthesia (halogens + morphine) [8].

Norepinephrine was chosen by Hong due to its ability to maintain systemic arterial resistance. Invasive blood pressure monitoring, pulmonary artery catheterization, and transesophageal ultrasound are among the recommended methods for proper monitoring.

This specialized equipment is specific to departments authorized to manage congenital heart disease. Pain stimulates the sympathetic system and causes a release of catecholamines, resulting in tachycardia and increased oxygen consumption. This can lead to cardiac decompensation, leading to cardiac arrest. Our analgesia was based on the use of step 1 and step 2 analgesics, combined with a nonsteroidal anti-inflammatory drug. It's also important to note that this is a relatively painless surgery on the artery wall.

CONCLUSION

Early detection and knowledge of the pathophysiology are essential for proper management. The key to anesthesia is, on the one hand, ensuring adequate monitoring to maintain a constant balance between peripheral and pulmonary vascular resistance and, on the other, preparing for the management of adverse events.

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Cite this article: Amadou HS, Mahamane YS, Hassane ML, Bachar LO, Alfred Dogbe YZ, Niandou M, Sabo R, Bandiaré M, Foumakoye A, Chaibou MS, (2025). Anesthesia in an Child with a Single Atrium: A Case Report and Review of the Literature. *EAS J Anesthesiol Crit Care, 7*(4), 72-74.