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Single Ventricle Type of Tricuspid Atresia with Pulmonary Stenosis in an 18-Month-Old Child in the Pediatrics Department of the Donka National Hospital/Guinea

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Abstract

Background: Univentricular hearts represent 0.5% - 1.5% of congenital heart defects. They are a heterogeneous group of malformations with the common feature of a single dominant ventricle. We report the case of an 18-month-old child with a single ventricle discovered following hospitalization for respiratory distress. **Objective:** To describe this rare and complex congenital heart disease and to draw practitioners' attention to the importance of antenatal screening in identifying fetal congenital heart disease early. **Case Presentation:** This was an 18-month-old child, hospitalized on January 12, 2025, for respiratory difficulty, cyanosis, and physical asthenia in the context of fever. Echocardiography-Doppler showed a single ventricle type of tricuspid atresia with pulmonary stenosis. **Conclusion:** Single ventricle remains a rare pathology and difficult to manage in our resource-limited countries.

Keywords

Ventricle, Single, 18-Month-Old Child, Pediatrics, Guinea

1. Introduction

The heart is the central organ of the circulatory system and is the engine of blood circulation. A longitudinal intracardiac septation subdivides the heart into two compartments, which are completely separated from each other after birth.

Cardiac embryogenesis is a very complex process requiring several stages, and

abnormalities can occur at each stage of development. Convergence abnormalities cause misalignment of the interatrial and interventricular septa, leading to complex heart diseases involving abnormalities in the development of the ventricles and atrioventricular valves. These heart defects can be grouped under the name "single ventricle".

However, Single ventricle is a rare congenital heart disease in which only one ventricular cavity can be identified, with a left morphology in 70% of cases.

It designates a pathophysiological concept where a single functional ventricle ensures pulmonary and systemic flow in parallel. This ventricle can be unique anatomically with the presence of a ventricle of normal morphology and another hypoplastic or atretic ventricle.

Biventricular repair consists of a cavopulmonary bypass, which allows the pulmonary and systemic circuits to be separated and a "serial" circulatory system to be reestablished Fontan-type circulation.

In addition to that, Prenatal diagnosis makes it possible to anticipate neonatal care [1]-[4].

We report the case of an 18-month-old infant with a single ventricle discovered following hospitalization for respiratory distress. Informed consent was obtained from the parents prior to the case description. The objective of our work is to describe this rare and complex congenital heart disease and to draw the attention of practitioners to the importance of antenatal screening to identify fetal congenital heart disease early.

2. Observation

It was an 18-month-old child, hospitalized on January 12, 2025 for respiratory difficulty, cyanosis, and physical asthenia, in the context of fever. The onset of the disease would go back a week marked by respiratory difficulty made of permanent polypnea of sudden onset, and cyanosis of the extremities. The parents consulted on January 7, 2025 in a local clinic where the child would have benefited from care based on undocumented injections. The evolution was marked by the persistence of respiratory difficulty and cyanosis with the appearance of fever and physical asthenia which led the parents to consult the child at the pediatric emergency room of Donka, and then they were transferred to admission for hospitalization and care.

Moreover, in his history, the child was born from an uneventful twin pregnancy, 6 Prenatal Consultations (CPN) were carried out, two (02) doses of tetanus vaccine (VAT), a serological assessment (negative retroviral serology, negative toxoplasmosis, negative rubella), and malaria chemoprophylaxis based on sulfadoxine-pyrimethamine and iron and folic acid supplementation received by the mother. The delivery was carried out in a health center by vaginal delivery, giving birth to full-term twins with an immediate cry and APGAR 10/10 at the 1st, 5th and 10th, with the following parameters: PN: 2500 g, T = 48 cm, PC: 31 cm. The newborns systematically received vitamin K1 and antiseptic eye drops. Breastfeeding was initiated early with mixed feeding from birth, currently on cereal and family

meals. Growth and development are below normal with delayed psychomotor development, vaccination status not up to date according to the expanded vaccination program of Guinea, with a history of recurrent cyanosis, the fifth child in a family of six, all alive and in good apparent health; born to a 30-year-old mother, a merchant, of Soussou ethnic group; no reported consanguinity, socioeconomic level is low.

Therefore, on physical examination, the weight was 6.6 kg; height was 82 cm, and weight/height index (W/H) between −2 and −3 Z-score, and a temperature of 37.8˚C with an unaltered general condition, asthenic, conscious, integuments and conjunctiva were colored, cyanosis of the extremities, tachycardia of 120 beats/min, systolic murmur 5/6 in the pulmonary focus; SpO2 = 80% AA, respiratory difficulty, tachypnea of 40 cycles/min, inter and subcostal indrawing; flaring of the nasal wings, crackling rales on pulmonary auscultation, there is digital clubbing, fine, dull, and brittle hair, the rest of the examination was strictly normal.

The biological assessment carried out in the emergency room, including blood count, hemoglobin electrophoresis, Hbs Antigen, hepatitis C serology, and retroviral serology, showed normality.

Chest X-ray showed a globular heart with bilateral pulmonary hypervascularization and increased bronchial network but no individual focus.

Doppler echocardiography showed a single ventricle of tricuspid atresia type with pulmonary stenosis.

Our patient received symptomatic treatment: oxygenation through the goggles at 3 l/min, an oral antianemic, infusible paracetamol 120 mg as needed (Figure 1).

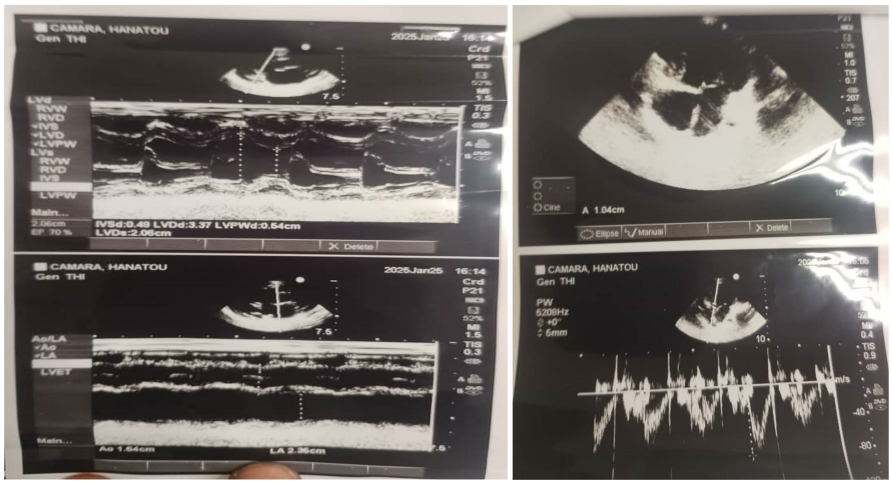

Figure 1. Excerpt from ultrasound images of the heart of the described case.

3. Discussion

Single ventricle is a rare congenital heart defect representing 0.5% to 1.5% of congenital heart diseases and appears in approximately 5 out of 100,000 newborns [2]. Kolié O. *et al.* recorded six cases of single ventricles over 4 years in the same

department but without pulmonary stenosis [5]. Our patient was female whose mother did not perform any prenatal ultrasound unlike Bedraoui in 2017 in Morocco who noted a male predominance, with a sex ratio G/F of 1.16 [4]. Our patient was 18 months old. In the study by Kolié O., the patients were between 0 and 2 years old, with an average age of 9.5 months [5]. This is consistent with the data in the literature where we find that most congenital heart diseases are diagnosed at an early age between 0 and 2 years. At the same time, Adehossi observed a patient who was diagnosed with univentricular heart at the age of 25 following an etiological search for polycythemia which would be due to the physiological reaction of the organism to tissue hypoxia [4]. The discovery of a single ventricle can be made from intrauterine life to adulthood, sometimes fortuitously. In our patient, respiratory difficulty, cyanosis and systolic murmur were the circumstances of discovery of the heart disease. This is consistent with the data in the literature, which shows that these signs are usual circumstances and indicative of a single ventricle in children. Cardiac Doppler ultrasound is the examination of choice for diagnosis. Symptomatic treatment was the only one that our patient benefited from due to the absence of cardiac surgery in our country. The surgical procedure aims to redirect the systemic venous flow directly to the pulmonary arteries, leaving the ventricle with the function of redistributing oxygenated blood within the systemic circulation; this is Fontan surgery [6]. Patient monitoring is carried out by pediatricians and adult cardiologists due to the absence of Guinean paediatric cardiologist. Since a single ventricle is a serious congenital heart disease, the mortality rate can reach 85% in the first two years of life; if the child survives the first two years, the mortality rate decreases to around 9% between 2 and 15 years of age. Indeed, the association of single ventricle and pulmonary stenosis would contribute to an increase in the lethality of these children. Some patients can reach 40 - 50 years of age or more depending on the type of associated malformations [2]. Moderate pulmonary valve stenoses are beneficial because they protect the pulmonary arterial bed and have a better prognosis [2] as is the case of our patient who has associated pulmonary stenosis. Over time, many patients who have benefited from Fontan surgery will develop irreversible heart failure requiring a heart transplant [7].

4. Conclusion

Single ventricle remains a rare and difficult-to-treat condition in resource-limited countries. Prenatal diagnosis is possible using ultrasound and allows for early neonatal care. But Surgical treatment remains palliative.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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