International Journal of Cardiology 184 (2015) 135–139

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Review

Successful pregnancy and delivery in patients with uncorrected single ventricle: Three new cases and literature review

Kana Wang [a], Hong Luo [b], Yalan Xin [c], Haiyan Yu [a,*]

[a] Department of Gynecology and Obstetrics, West China Second Hospital, Sichuan University, Chengdu, Sichuan 610041, China
[b] Department of Diagnostic Ultrasound, West China Second Hospital, Sichuan University, Chengdu, Sichuan 610041, China
[c] Department of Gynecology and Obstetrics, Meishan City Women and Child Care Hospital, Meishan, Sichuan 620010, China

ARTICLE INFO

Article history:
Received 12 July 2014
Received in revised form 14 November 2014
Accepted 25 January 2015
Available online 27 January 2015

Keywords:
Univentricular heart
Pregnancy
Risk factor
Prognosis

ABSTRACT

Due to high risks of both maternal and fetal complications, pregnancy is not encouraged for women with uncorrected univentricular heart (UVH). Here, we report three cases of successful pregnancy and delivery in patients with uncorrected UVH. A literature review has been performed. It appears that maternal and neonatal risks are mainly associated with higher NYHA heart failure class, pulmonary hypertension, and history of congestive heart failure. In the absence of these risk factors, successful pregnancy still can be achieved with mild complications. Care by a multidisciplinary team during delivery is necessary to for a good prognosis.

1. Introduction

The occurrence of univentricular heart (UVH, or single ventricle) is rare, accounting for only 0.5–1.5% among patients suffering from congenital heart disease (CHD) [1,2]. In such condition, single ventricular pumping chamber supplies both systemic and pulmonary circulations, in which oxygenated and deoxygenated blood are mixed. Patients usually exhibit certain degree of systemic arterial desaturation [3]. Survival beyond childhood and early adolescence with uncorrected single ventricle is unusual. Therefore, pregnancy under this condition is rarely successful. In 1963, Mandel & Hirsch [4] first reported a pregnancy case in a 29-year-old woman, who died post-partum. To date, less than 20 cases of successful pregnancy with uncorrected UVH have been reported. In this article, we report three cases of successful pregnancy and delivery with uncorrected UVH. In addition, we performed a Medline search and reviewing for English-language literature for the similar cases in the past 15 years, which are summarized in the current study.

1.1. Case 1

A 26-year-old patient (153 cm, 54 kg; gravida 3, para 0) was diagnosed for UVH with double inlets by cardiac ultrasonography when she was 16-year old. Surgical treatment was proposed but declined by her parents. Fortunately, her cardiac function was in New York Heart Association Class I (NYHA I). Before the current pregnancy, she was in a stable condition without excise intolerance. Her obstetrical history included a spontaneous abortion 5 years ago and an induced abortion 4 years ago. Because of the high risk associated with continued pregnancy, medical abortion was indicated in the first trimester of this pregnancy but refused by the couple. In the first trimester, the patient was in a stable condition without exercise intolerance and manual work limit. In the second trimester, her exercise intolerance was gradually increased. Fetal detailed ultrasound scans were performed at 28 and 32 weeks. The fetus showed a normal biophysical profile without cardiac anomaly. The umbilical artery blood flow S/D ratio was 5.5. At 32 weeks, the patient started to experience fatigue and exertional dyspnea. However, the estimated fetal weight was less than the 10th percentile in both scans, indicating small for gestational age (SGA) [5]. At 34 weeks, she was admitted to our hospital due to false labor and bloody show.

Physical examination upon admission revealed cyanosis on lips, ear lobes and nail beds, as well as marked clubbing of the fingers and toes. Her pulse was regular and the rate was 75 beats/min (bpm). There was no discrepancy between pulse and heart rate (HR). Her blood pressure was 139/89 mm Hg, and oxygen saturation by pulse oximetry (SpO_2) was 80%. Dynamic electrocardiogram (DECG) monitoring and oxygen mask were immediately applied. Her SpO_2 increased to 91%. The external jugular venous (EJV) pressure was normal. Blood test showed RBC: 4.61×10^9/l, hemoglobin (Hb): 15.2 g/dl, Hct: 46.7%, and platelet: 108,000/ml. Gas analyses showed normal results. Echocardiography revealed a complex malformation consisting of (1) single ventricle (left type) with double outlets with an ejection fraction (EF) of 58%,

* Corresponding author.
E-mail address: fanjy422@163.com (H. Yu).

http://dx.doi.org/10.1016/j.ijcard.2015.01.052
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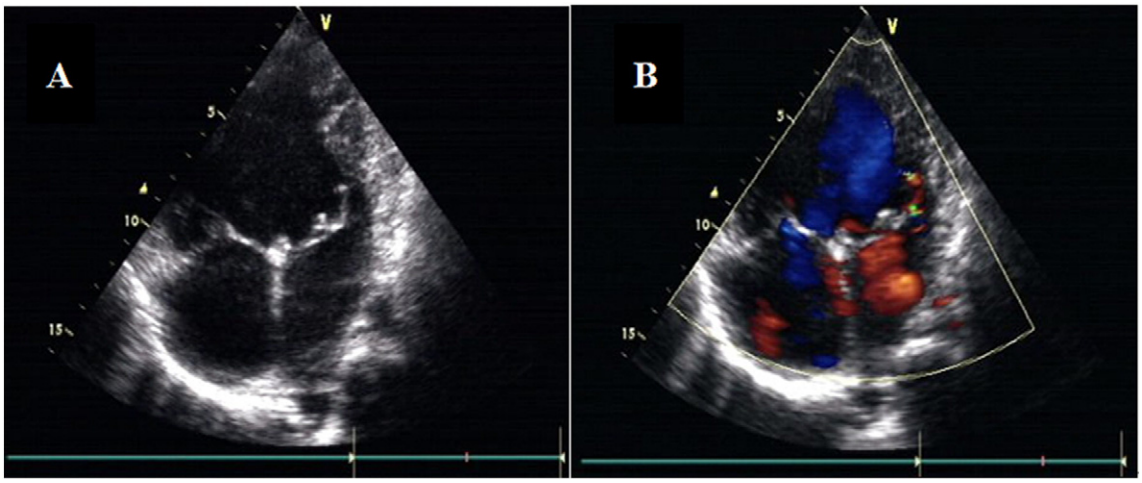

Fig. 1. Echocardiography of patient in Case 1. (A) Single ventricle (left type) with double outlets (EF = 58%, apical four-chamber view). (B) Blood flow in early systole (apical four-chamber view).

(2) severe regurgitation in pulmonic valves, (3) dextro-malpositions of aortic arch, and (4) moderate regurgitation in tricuspid and mitral valves, but with normal veno-atrial connection. The ECG showed a sinus rhythm with extreme right axis deviation (Fig.1 and Fig. 2) (+105°) and T-wave alternants.

On the next day of admission, caesarean section (C-section) was performed with the support of a multidisciplinary team (cardiologist, thoracic surgeon, neonatologist, and anesthetist) under combined spinal–epidural anesthesia. A 1330 g female infant was successfully delivered with Apgar scores of 8, 9 and 10 at 1, 5 and 10 min, respectively. The patient had approximately 400 ml blood loss but maintained stable vital signs. After childbirth, the patient received prophylactic *Cefathiamidine*. Pain relief was achieved with *Tramadol* through patient-controlled analgesia (PCA) pump. The patient recovered uneventfully and was discharged home at 7 days after operation. Congenital cardiac defect was not detected in the infant. Three years later, the patient was in a good condition and the baby was growing as expected.

1.2. Case 2

A 20-year-old primigravida was confirmed to have UVH in the current pregnancy. She was diagnosed with congenital heart disease at an early age but did not receive a diagnosis for the type of defect because she had no symptom and never visited a cardiology specialist. She remained asymptomatic until the second trimester. At 33-week gestation, she developed paroxysmal nocturnal dyspnea and orthopnea, which can be relieved by sitting straight. At 34 weeks and two days of gestation, the patient was admitted to our hospital because of premature rupture of membranes.

Upon admission, she had a BP of 116/71 mm Hg, HR of 116 bpm, and SpO_2 of 86% (with oxygen mask). Blood test showed RBC: 4.94×10^9/l, Hb: 16.2 g/dl, Hct level: 49.3%, and platelet: 121,000/ml. Cyanosis and marked clubbing of the fingers and toes were present. Gas analyses showed normal results. There was a grade IV systolic murmur at the heart area. The EJV pressure was normal. Her cardiac function was

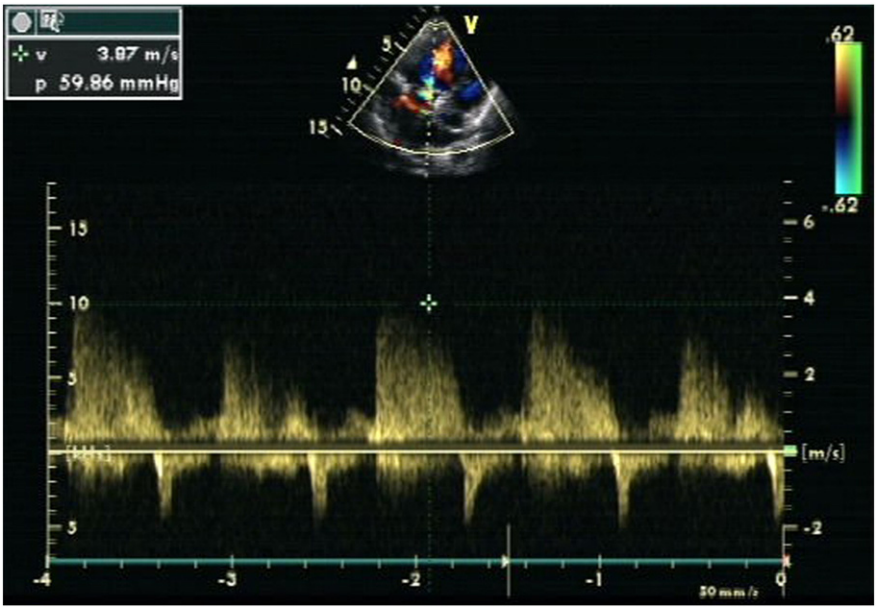

Fig. 2. Continuous wave Doppler echocardiography in Case 1. Severe regurgitation in pulmonic valves, and moderate regurgitation in tricuspid and mitral valves.