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Case Report

Pregnancy and delivery in a patient with a Fontan circulation and primary ciliary dyskinesia: A case report

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ABSTRACT

A patient had primary ciliary dyskinesia with a complex cardiac malformation. As a child, she had benefited from a Fontan surgery to maintain a proper cardiac function. In such patients, whether it is safe to become pregnant is controversial. This case illustrates the possibility of carrying a pregnancy to term and providing a vaginal birth if a rigorous preconception consultation is performed to ensure care by a multidisciplinary specialized team, and the patient is properly informed of the risks.

Case report

The patient has been followed since birth for a complex cyanogenic heart disease. She initially was diagnosed with heterotaxy (association of a complete situs inversus with a complex cardiac and vascular malformation). In addition to dextrocardia, the heart exhibited a single ventricle, a stenosis of the pulmonary ejection pathway, and an absence of resorption of the inferior vena cava (located on the left hand side of the column). Moreover, there were two superior vena cava and blood drained into the right atrium through the coronary sinus. The pulmonary venous system was also aberrant, since it was shared between the two atria. The mitral valve was atretic with blood flowing through an interatrial communication from the left atrium to the right. Finally, after passing through a left tricuspid valve, the blood reached a stenosed pulmonary ejection pathway and the aorta. (Fig. 1) This malformation resulted in a lack of perfusion of the pulmonary circulation causing inefficient oxygenation . This was well tolerated until the age of 3 years when cyanosis increased.

The patient then underwent a first palliative surgery during which pulmonary perfusion was improved by inserting a shunt between the subclavian artery and the right pulmonary artery (Blalock operation) through a Gore-Tex tube. At the age of 8 years, Fontan surgery modified according to de Leval's procedure was performed to improve cardiac performance and prevent the tricuspid valve from becoming progressively insufficient. Perfusion in the pulmonary arteries was increased by anastomosis of the superior homolateral vena cava, and the interatrial communication widened to improve the flow of pulmonary blood from right to left. Finally, the blood of the inferior vena cava was entirely deviated from the pulmonary trunk. The final blood circuit consisted of a heart with two in series circulations; the blood returning from the peripheral venous system progressed in the pulmonary circuit with no cardiac contraction thanks to its residual inertia (Fig. 2).

Primary ciliary dyskinesia (PCD) was diagnosed when the patient was 14 years old because of, situs inversus and repeated respiratory infections during childhood, for which symptomatic treatment was started (respiratory physiotherapy, oral azithromycin prophylaxis and corticosteroid nasal sprays). At 29, the patient considering pregnancy, a full cardiologic assessment was performed. Clinically, the patient had no limitations in everyday life activities (NYHA class 1). Cardiac ultrasound showed good single ventricular function with an ejection fraction (EF) of 64%. Electrocardiogram (ECG) identified an axial deviation and atypical morphology of the QRS complex in relation to the underlying cardiopathy. The sinus rhythm was subnormal, presenting some polymorphic ventricular extrasystole. The spirometry tests demonstrated a satisfactory functional capacity. No

Abbreviations: PCD, primary ciliary dyskinesia; EF, ejection fraction; ECG, Electrocardiogram; PPH, postpartum hemorrhage; LMWH, low-molecular-weight heparin

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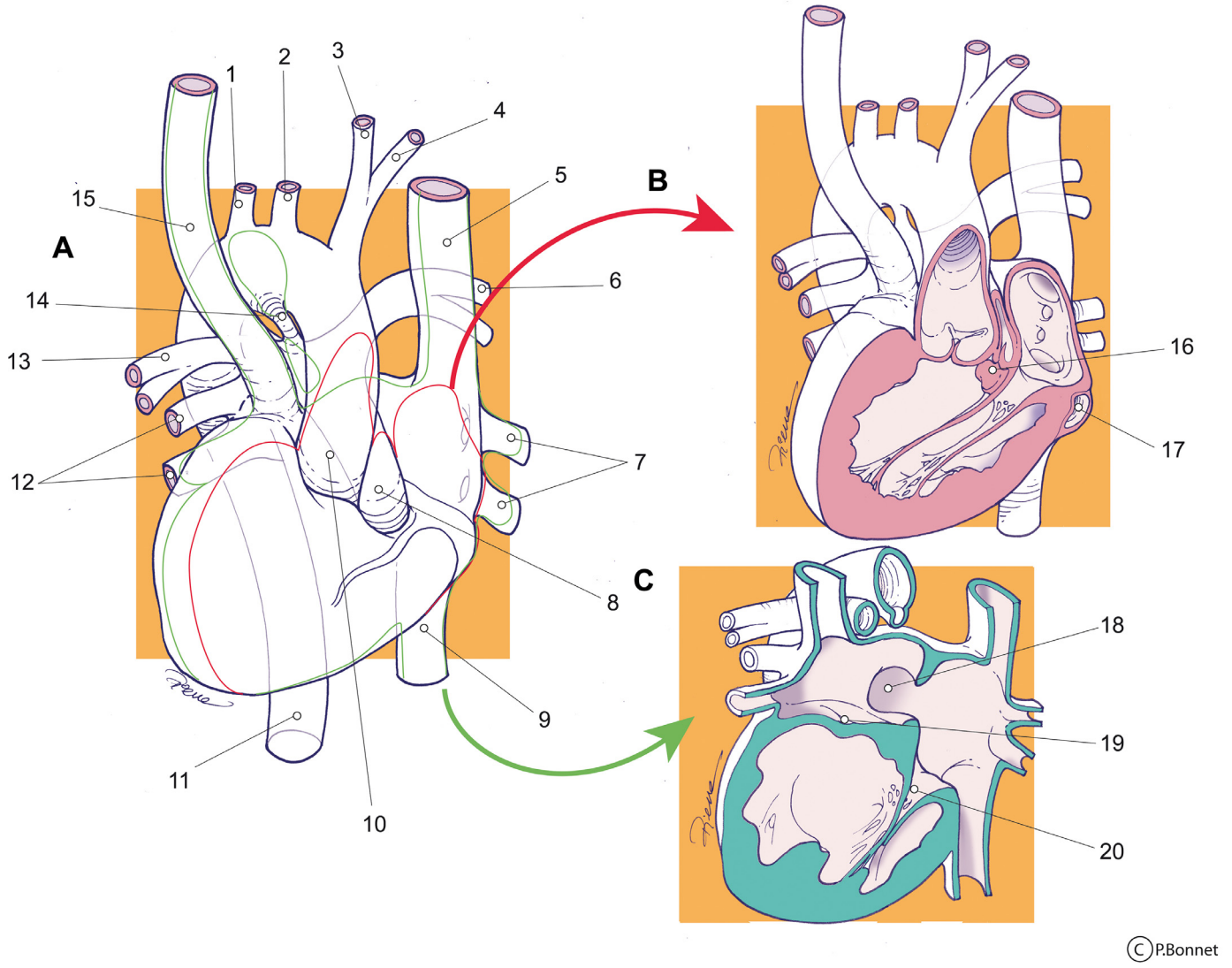

Fig. 1. Heart malformation. (1) Right subclavian artery (2) Right common carotid artery (3) Left common carotid artery (4) Left subclavian vein (5) Left superior vena cava (6) Left pulmonary artery (7) Left pulmonary vein (8) Pulmonary trunk (9) Inferior cava vena (10) Aortic bulb (11) Ascending aorta (12) Right pulmonary vein (13) Right pulmonary artery (14) ductus arteriosus (15) Right superior vena cava (16) pulmonary valve (17) Coronary sinus (18) interatrial communication (19) Mitral valve (20) tricuspid valve.

supraventricular arrhythmias were found using a Holter monitor. Moreover, respiratory function tests were within the normal range.

Preconception counselling was carried out to ensure that the patient understood the cardiological and obstetrical risks of a pregnancy with this condition. Because of the PCD and a history of salpingectomy, in vitro fertilization was carried out, leading to an intrauterine singleton pregnancy. No PCD related mutation was found avoiding the realization of a preimplantation genetic diagnosis.

Pregnancy was uneventful on an obstetrical point of view. The patient suspended her professional activities from the beginning of pregnancy. The patient's condition remained excellent throughout the pregnancy. There was no sign of cardiac decompensation. Quarterly cardiac assessment were performed. ECGs showed uncomplicated isolated ventricular extrasystoles with no clinical repercussions and no supraventricular arrhythmia while concomitant maternal hearth ultrasound demonstrated a stable, correct ventricular function. Fetal monitoring was reassuring with normal morphology scanning ultrasound, especially from a cardiac point of view. Pulmonary-wise, the patient benefited from physiotherapy and daily treatment with a corticosteroid nasal spray, as well as chronic anti-infection prophylaxis with azithromycin. The patient remained eupneic, and the bronchorrhea associated with PCD was stable. Microbiological and sputum analyses were regular. The quarterly spirometry tests were within normal range throughout the pregnancy.

At 37 weeks of pregnancy, the patient was hospitalized in the high-risk pregnancy department to manage delivery. Labour induction at 38 weeks of pregnancy was decided because of the cardiac pathology. Because of an unfavorable Bishop score, Cook© balloon was used. Continuous epidural analgesia was administered as soon as the patient was in labor. Continuous hemodynamic monitoring was performed by noninvasive monitoring of blood pressure and cardiac output. The labor was carried out without complications. Midforceps delivery allowed the birth of a healthy boy as soon as the fetal head was engaged. Neonatal adaptation was excellent (birth weight of 2802 g and Apgar score of 8/9/9). Immediate postpartum hemorrhage (PPH) required manual delivery and carbetocin administration. Monitoring in the postpartum period was not complicated by any hemodynamic dysfunction. Antithrombotic prophylaxis with low-molecular-weight heparin (LMWH) was initiated. The patient left the hospital on the fifth postpartum day. Patient cardiological follow up during the subsequent year did not demonstrate any deterioration in his hemodynamic status.

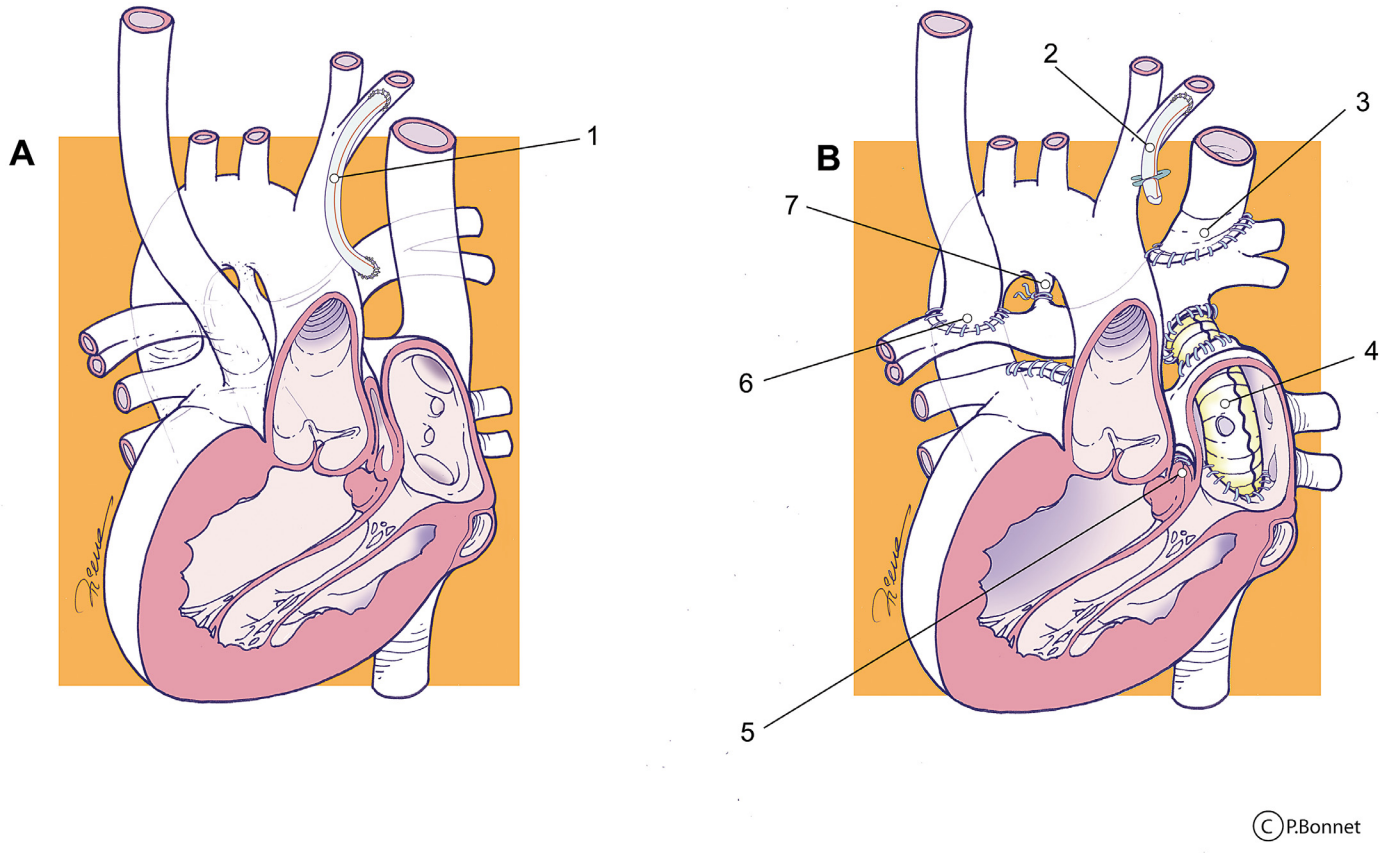

Fig. 2. Heart Reparation. Blalock surgery: (1) Blood from subclavian artery redirected to the pulmonary artery, de Leval modified Fontan surgery: (2) Blalock shunt occlusion (3) Right superior cava vena to right pulmonary artery anastomosis (4) Inferior cava vena to right pulmonary artery anastomosis (through a intra auricular shunt) (5) Pulmonary Valve (6) Left superior cava vena to left pulmonary artery anastomosis (7) Ductus arteriosus ligation.

Discussion

1). Primary ciliary dyskinesia

PCD is a genetic condition that causes defects in the cilia function. Respiratory complications constitute the essential part of its clinical expression. Because of the relative scarcity of PCD, treatments are extrapolated from those for chronic respiratory diseases, such as cystic fibrosis [1].

Data from the literature are reassuring about morbidity and mortality during pregnancy. The two limiting factors in terms of the maternal prognosis are a maximal expiration volume per second less than 50% of the theoretical value and the presence of multiresistant organisms [2]. Concerning the fetal prognosis, the preterm birth rate is increased compared to the general population. PCD does not contraindicate a vaginal delivery; assisted delivery can help to limit the active expulsion time.

2). Situs inversus and Fontan circulation

The clinical impact of a complete, isolated situs inversus is rare. Obstetrically, this anomaly must be known and taken into consideration in the prevention and treatment of vena cava syndrome. Nevertheless, the absence of a double heart pump leads to a more fragile hemodynamics that depends on systemic venous pressure for the oxygenation process [3]. In addition, the structural and functional cardiac modifications expose these patients to supraventricular arrhythmias (with an incidence up to 40% by 10 years after surgery) [4].

Venous and cardiac embolism are other long-term side effects of this surgery. Finally, systemic venous hypertension may lead to protein-losing enteropathy [5,6]. Despite surgical advances, mortality at age 20 still reaches 15% [7]. Nevertheless, many patients reach the age of childbearing.

a) *Maternal hemodynamics*

These physiological changes consist in a 10–20% increase in the heart rate, a 40% increase in blood volume, and a 30–50% increase in cardiac flow. The labor and the postpartum period are particularly at risk of cardiac decompensation [8]. It is therefore essential to carefully evaluate these patients to try to identify those for whom the cardiovascular risk associated with pregnancy is acceptable.

Maternal cardiovascular complications reported in this population are supraventricular arrhythmias, cardiac decompensation, chest pain, thromboembolic events, and hypoxia [9]. Several criteria are used to stratify and establish a risk score in patients with congenital or acquired heart disease during pregnancy. The two most frequently used score are CARPREG [10] and the modified WHO score (mWHOs) [11].

Preconception counselling

The issue of recommending or discouraging pregnancy in a patient with a Fontan circulation is still under debate [12–15]. However, it is essential to carry out a rigorous preconception assessment before considering pregnancy. Echocardiogram, ECG, cardiac MRI, exercise stress test can be useful in this regards [16,17].

a) *Obstetrical and neonatal management*

In these patients, fertility appears to be lower. The hypothesis of chronic hypoxemia and ovarian venous congestion has been

suggested. However, the classically unfavorable opinion of gynecologists concerning pregnancy may have caused bias in these studies. In case of assisted reproduction technology in these fragile patients, precautions will be taken for reduce the risk of thromboembolic complications, ovarian hyperstimulation syndrome and multiple pregnancies.

A higher rate of miscarriage (three times more than the general population) has been established [13]. Previous literature has reported higher incidences of premature deliveries, premature rupture of membranes, and PPH (up to 50%) [18]. This is probably related to the fear of using uterotonics due to their cardiovascular effects, as well as the large proportion of patients receiving thromboprophylaxis with LMWH and/or aspirin. The frequent uses of instrumental extraction and cesarian section are also contributing factors, but Fontan circulation is an independent risk factor for an increased incidence of PPH [19, 20].

Finally, a higher rate of cardiac malformations is expected and a specialist second-line ultrasound is recommended.

a) *Mode of delivery*

In the absence of obstetric contraindication or major heart failure, vaginal delivery should be encouraged. Instrumental extraction must be performed to avoid the decrease in systemic venous return induced by Valsalva's efforts [10,12–14].

a) *Anesthetic management*

As in the vast majority of heart diseases, regional anesthesia for vaginal delivery represents a medical indication. It contributes to minimize the hemodynamic changes related to labor and to reduce the levels of circulating catecholamines [18]. In case of cesarian section, a neuraxial technique with catheter allows for progressive titration of anesthetics avoiding a sudden decrease of systemic vascular resistances and venous return. General anesthesia should be avoided because, in addition to all its limitations in the obstetric context, it exposes patients to a risk of major hemodynamic alterations, mechanical positive pressure ventilation leading to an increased intrathoracic pressure and a subsequent decrease in an already compromised systemic venous return [3].

a) *Thromboprophylaxis*

A Fontan circulation represents a situation with high thromboembolic risk linked to the passive blood flow in the pulmonary circulation and to the presence of prosthetic material. The prevalence of intracardiac thrombi can reach 20%. Systematic antithrombotic prophylaxis remains controversial, but it should be discussed based on the patient's history [5,6].

In conclusion, pregnancies in patients with a Fontan circulation are rare, but their management must be attentive. The association with a PCD is exceptional. Our case report and some series in the literature show that it is possible to achieve a pregnancy under these conditions. However, a multidisciplinary preconception evaluation is needed. A complete and objective information should be provided to the patient about medical, obstetric, and neonatal risks. Multidisciplinary care is essential during all follow up. It is necessary to anticipate well-known maternal complications, such as arrhythmias or postpartum hemorrhages. Adequate understanding of the physiological alterations of the cardiovascular system during pregnancy, labor, and delivery and their effects on this modified cardiovascular circulation allows for the provision of optimal care to the mother and newborn.

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Declaration of Competing Interest

The authors report no conflict of interest.

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