



## Favourable Outcome of Two Contraindicated Pregnancies in a Woman with Complex Congenital Heart Disease and Eisenmenger Syndrome

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### Abstract

We report the case of a young woman who was born with complex heart disease consisting in a single ventricle, double discordance, tricuspid atresia without pulmonary obstacle, and fixed Pulmonary Arterial Hypertension (PAH). She carried two pregnancies to term against medical advice after percutaneous dilatation of a restrictive atrial septal defect.

The good functional tolerance of her heart disease (class I-II NYHA), saturation greater than 85%, and early multi-disciplinary management of the pregnancies undoubtedly contributed to their successful outcome.

Our patient's experience should not call into question the contraindication for pregnancy in this type of disease but may help in the management of similar women who, although aware of the risk, desire to become pregnant.

### Introduction

The prognosis of Eisenmenger Syndrome (ES) is usually better than that of idiopathic Pulmonary Hypertension (PAH) [1]. However, the more complex the underlying heart disease, the less favourable it becomes [2]. Pregnancy carries a high risk of mortality for women with ES and is therefore strictly contraindicated [3-6].

### Case Presentation

We report the case of a young woman who was born with complex heart disease consisting in a single ventricle, double discordance, tricuspid atresia without pulmonary obstacle, and fixed Pulmonary Arterial Hypertension (PAH) who became pregnant against medical advice and carried two uneventful pregnancies successfully to term.

Mrs P. was born 3/4/1980. The precise diagnosis of her heart condition was made by ultrasonography when she was 8 years old following pneumonia. Her only extra-cardiac history was scoliosis, which was operated on under loco-regional anaesthesia at the age of 14 years. Her clinical status remained stable throughout adolescence with stage III dyspnoea, saturation between 75 and 80% and polycythemia (haemoglobin level of 20.5 g/l), and she was able to work part-time as a secretary. In November 2004, she was admitted to hospital for worsening of her dyspnoea during a period of bronchial superinfection. A diagnosis of pulmonary oedema was suspected on the basis of combined cough, orthopnoea and disclosure of bilateral crepitations. Chest x-ray showed diffuse interstitial syndrome, and ultrasonography restrictive Atrial Septal Defect (ASD) (Figure 1). The patient improved on diuretics and antibiotics.

It was decided to dilate her ASD by interventional catheterism under Transoesophageal Echocardiographic (TEE) guidance. TEE confirmed diagnosis, showing spontaneous contrast in the left atrium, which was very "stretched" and measuring a continuous gradient through the ASD of 16 mmHg. PAH was systemic (Table 1) with elevated capillary pressure. The ASD was dilated with a balloon 25 mm in diameter (Figure 2). At the end of the procedure, left atrial pressure fell to the level of that of the right atrium but PAH remained systemic (Table 1, Figures 3 and 4).

The patient's functional improvement was dramatic (class I-II) and she was able to walk up four flights of stairs, and put on weight (+7kg). She began considering the possibility of conceiving a

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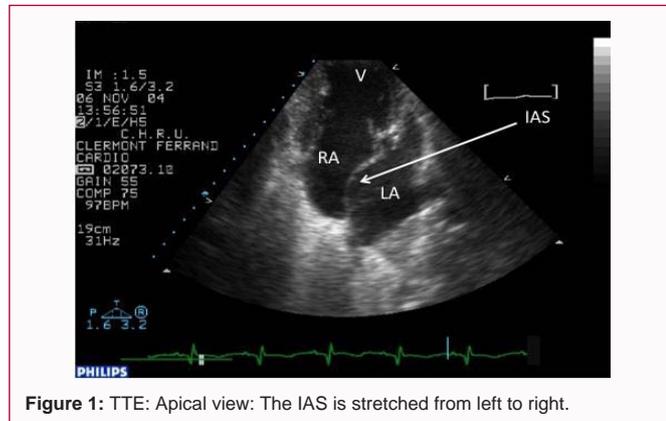
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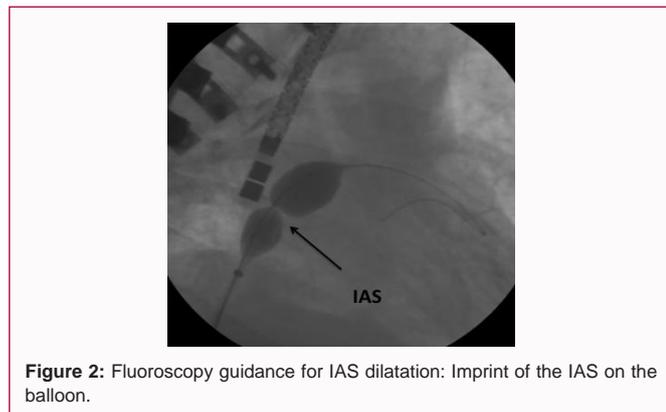
**Table 1:** PAH was systemic with elevated capillary pressure.

	RAP	LAP	PAP (S/D m)	Aortic pressure
Pre-procedure	8	26	81/44 m 59	75/50 m 50
Post-procedure	10	14	73/35 m 52	

RAP: Right Atrial Pressure; LAP, PAP: Pulmonary Artery Pression (S: Systolic, D: Diastolic and M: Moyenne).



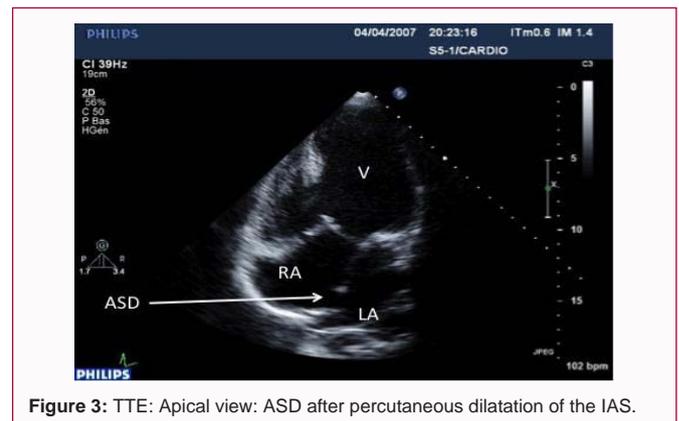
**Figure 1:** TTE: Apical view: The IAS is stretched from left to right.



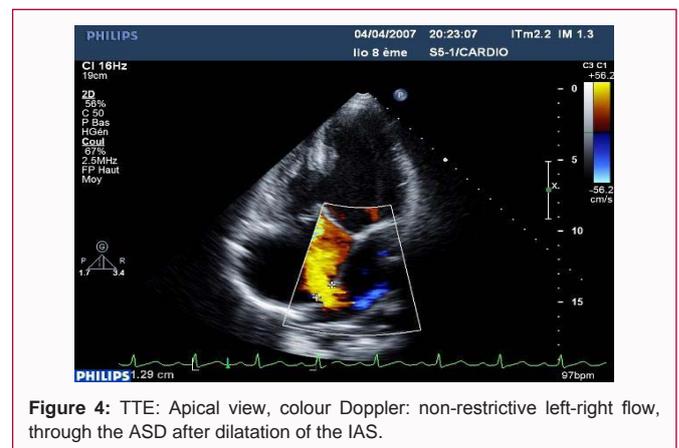
**Figure 2:** Fluoroscopy guidance for IAS dilatation: Imprint of the IAS on the balloon.

child. Saturation was 85%.

Despite repeated reminders of the contraindications, Mrs. P became pregnant. She presented in September 2006 in her seventh week of pregnancy with saturation 92% and a haemoglobin level of 16.5 g/dl. The couple were once again warned that the pregnancy exposed mother and unborn child to risks but they decided nevertheless to go ahead. A cardiological examination was performed every 4 to 6 weeks and a multidisciplinary team comprising anaesthetists, cardiologists and obstetricians discussed how the birth should be managed in the event of an emergency. All decisions arrived at were recorded in the patient's file. The patient's pelvis was narrow and so it was decided to perform a caesarian section, before term but if possible after 32 weeks, under continuous spinal anaesthesia with invasive monitoring of blood pressure. During pregnancy, her clinical status was stable with saturation greater than 85% and she gave birth to a boy weighing 1 kg 945 (APGAR 8-10-10) at 34wk+4d. She was monitored for 48h in the cardiology intensive care unit and then 5 days in the maternity unit. The child stayed 1 month in the neonatology department. Treatment with Low-Molecular-Weight Heparin (LMWH) at a curative dose was prescribed, later replaced by Anti-Vitamin K (AVK). Diuretics were reintroduced distance to birth owing to left- and right-congestive signs, and Converting Enzyme Inhibitor (CEI) treatment was initiated in November 2008 of systematic way (single ventricle



**Figure 3:** TTE: Apical view: ASD after percutaneous dilatation of the IAS.



**Figure 4:** TTE: Apical view, colour Doppler: non-restrictive left-right flow, through the ASD after dilatation of the IAS.

and slight leakage from the right mitral valve).

Mrs. P. became pregnant a second time. She presented in February 2012 at 5 weeks of amenorrhoea with the firm intention of continuing the pregnancy despite reiteration of the risks involved. On her own initiative, she stopped CEI treatment. AVK were replaced by LMWH during the first term. The pregnancy was managed under the same conditions as the first. She underwent caesarian section at 35 weeks of amenorrhoea after corticosteroid treatment to accelerate fetal lung maturation and replacement of AVK by LMWH, under continuous spinal anaesthesia. At the patient's request, perioperative tubal ligation was performed. She gave birth to a girl weighing 2 kg 030 (APGAR 3, 6, 8), who was admitted to paediatric intensive care for respiratory distress. Postpartum progress was uneventful except for marked asthenia, which delayed discharge until day 16.

## Discussion

In recent years, the number of adults suffering from complex congenital heart disease has continued to rise owing to improved management of the condition. Marelli [7] reported a prevalence of 0.38 per1000 adults, with a female predominance (57%). In developed countries, maternal heart disease has become the leading cause of death during pregnancy [8], and congenital heart disease the most frequently encountered heart disease in pregnant women [9].

Such patients should be monitored at least once a year at a "GUCH" specialised centre [4]. In the case of our patient, this follow-up led to a reassessment of her condition, which until then had been considered as stable and intractable to treatment, and the decision to perform percutaneous dilatation of her atrial septum, which although only a palliative intervention transformed her functional status. The

recommendations concerning pregnancy in patients with a single ventricle are clear: "Pregnancy is contraindicated in cases of severely restricted pulmonary airflow, severe pulmonary vascular disease (Eisenmenger syndrome) and poor ventricular function" [4]. The only element in our patient's favour before pregnancy was her class II NYHA [3].

PAH is an absolute contraindication for pregnancy, with a reported maternal mortality rate of 30 to 50% in earlier series [10], and one of 17 to 33% in more recent publications [11]. Only patients with positive long-term response to calcium channel blockers seem to have a more favourable prognosis [12]. The risk of maternal death occurs during the last trimester of pregnancy or in the first weeks postpartum owing to PAH attack, pulmonary thrombosis or intractable right heart failure. Eisenmenger syndrome [3-5] compounds the risks of PAH and right-left shunt, which can worsen during pregnancy as the result of decreased systemic vascular resistance. A recent publication reports a cardiac morbidity of 33% and a mortality of 5% in a French multi-center series of 20 patients (28 pregnancies) [13]. If the pregnancy is continued, rest, anticoagulation therapy, (at least preventive regimen) and controlled intake of iron substitutes are recommended [14]. Oxygen saturation and blood counts should be regularly monitored. Owing to haemodilution, the level of haemoglobin is less reliable at the end of pregnancy than saturation [15].

Maternal cyanosis is itself a serious risk factor during pregnancy, in particular for the fetus. In a series of 96 pregnancies in 44 patients with cyanotic heart disease, excluding those with Eisenmenger syndrome, Presbitero et al. [16] reported a maternal morbidity rate of 32% and only one death but a high rate of fetal complications: premature birth (37%), hypotrophy, miscarriage, and fetal death in utero. The percentage of live births was directly related to the extent of hypoxia (12% of live births when saturation was lower than 85%, 45% from 85 to 90%, and 92% when it was higher than or equal to 90%), to the level of haemoglobin (71% of live births when it was lower than or equal to 16 g/dl, 45% between 17 and 19 g/dl and 8% when it was higher than or equal to 20 g/dl), and to the type of heart disease (31% of live births in the "single ventricle" group).

Cardiac failure can be prevented by rest and early administration of diuretics, at the lowest dose possible to avoid haemoconcentration. Because of the occurrence of endocarditis postpartum in 7% of the patients (one of whom died 2 months after delivery), the authors consider prophylactic antibiotic treatment to be mandatory during delivery.

Management of delivery [6] should be discussed for each individual case according to the patient and the practice of the medical team. Swan Ganz catheterisation is not advisable since the risk of complications outweighs the potential benefits [3]. The risks of becoming pregnant should be explained early on to young women with severe heart disease, re-explained at regular intervals and once again in the event of pregnancy. Termination of the pregnancy can be envisaged because of the threat to the mother's health but itself is not without risk [6].

In their article Kovacs et al. [15] questioned women with congenital heart disease to see whether their physician had informed them of the risks involved in becoming pregnant. Of the 80 women at intermediate to high risk, 34 % had received no information, and out of 18 women for whom pregnancy was contraindicated, 9 had

not been warned of the potential harm. Presbitero et al. [16] observed functional impairment in their patient series during the 2 years following delivery but were unable to conclude whether it was due to the natural progress of the disease or to the additional workload of looking after an infant. This possibility should be discussed with the couple before or during the pregnancy and social assistance should be systematically offered during the postpartum period.

## Conclusion

Improved management of congenital heart disease has led to an increase in the number of women of childbearing age suffering from the condition. Cyanotic heart disease, particularly in combination with PAH, such as Eisenmenger syndrome, still remains a contraindication for pregnancy because of the high risk of maternal mortality. Although informed of the risk to life involved in becoming pregnant, certain women decide nevertheless to try to conceive. Early and multidisciplinary management of the pregnancy in a specialized center should be undertaken to ensure mother and child every chance of successful outcome.

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