Successful pregnancy after pulmonary artery banding; case report

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Figure 1
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Abstract

Although, double inlet left ventricle is a rare congenital heart defect, it is still treated by pulmonary artery banding in developing countries.

We report a successful pregnancy outcome in a patient with pulmonary artery banding. Patient condition worsened by 34 weeks gestation and she developed sever reduction of diastolic blood flow. Cesarean section was done for her and she give birth to a boy observed in NICU for two weeks and both were discharged in good condition.

It is possible for patients with pulmonary artery banding for double inlet left ventricle to carry on pregnancy but with vigilant and close follow up.

Introduction

Pulmonary artery banding (PAB) was introduced by Muller and Danimann in 1951 as technique of palliative surgical therapy used as a staged approach to operative correction of congenital heart defects [1]. This technique was widely used in the past as an initial surgical intervention for infants born with cardiac defects characterized by left-to-right shunting and pulmonary overcirculation. Within the last two decades, early definitive intracardiac repair has largely replaced palliation with pulmonary artery banding. The primary objective of performing pulmonary artery banding is to reduce excessive pulmonary blood flow and protect the pulmonary vasculature from hypertrophy and irreversible (fixed) pulmonary hypertension [1-3]. Patients who have single ventricle defects have the aorta arises from an outflow chamber [4, 5]. PAB is indicated operation for single ventricle heart disease which has incidence of 5 cases per 100 000 live births [1, 3].

The principle of PAB is the reduction of the diameter of the main pulmonary artery, decreasing blood flow to the pulmonary branches and reducing pulmonary artery pressure. Concomitant improvement of systemic pressure, cardiac output and ventricular function [3, 6]. At the same time, PAB exposes the patient to the risk of low systemic oxygen saturation as a consequence of unbalanced mixing between the systemic and pulmonary venous blood, or abnormal ventricular hypertrophy, or sub-aortic obstruction as well as various degrees of pulmonary branch and valve distortion [2, 3, 6].

Despite serious long-term sequelae, such as impaired systemic ventricular function and systemic atrioventricular valve regurgitation, patients have reached and will reach childbearing age [1]. Pregnancy itself encompasses several important haemodynamic changes (for example, increased cardiac output and reduced systemic vascular resistance) that may potentially threaten the health of both the mother with PAB and her offspring [2, 3]. In particular, the surplus of systemic venous return may lead to complications, such as atrial arrhythmias, oedema and ascites. On the basis of the available literature, the pre-existent routine tendency to discourage pregnancy was abandoned [5, 6]. The presence of long term sequelae after PAB and the need for drugs (for example, oral anticoagulation) were suggested as pregnancy risk indicators [2, 3]. Here, we report a successful pregnancy in a woman with PAB.

Case report

This is a 30 years old Saudi lady primigravida admitted to ICU as G1 P0+0 known case of Complete cyanotic cardiac disease. At age of 6 years after cardiac catheterization for shortness of breath (SOB), and cyanosis on exertion to have double inlet left ventricle, Hypoplastic right ventricle and transposition of great arteries. She had pulmonary artery banding done followed by recatheterization. Her mitral valve area (MVA) pressure was 40/15 with mean pressure of 26. PA gradient of 80mmHg and she had cardio pulmonary anastomosis for elevated pulmonary artery pressure (PAP).

When she came to emergency room (ER), she was 26 weeks and complaining of SOB, left leg swelling and exertion for the past 6 weeks. Her oxygen saturation was 91% and she was evaluated and followed by cardiologist. On examination, she was having palpable S1, left parasternal heave, pan systolic murmur all
over the chest, and gallop rhythm. Patient improved and was discharged on Metoprolol, Frusemide, and Clexan. She was followed in the outpatient department (OPD).

At 33 weeks gestation, patient was discovered to have reduced diastolic flow on Doppler ultrasound. She was advised to follow up weekly in the OPD clinic and planned for elective caesarian section at 37 weeks. Within five days patient presented to ER complaining of severe SOB and was admitted to ICU. On examination, vital signs were normal, with early systolic murmur, mild cyanosis, clubbing, and mild respiratory distress. ECG showed normal sinus rhythm. Echo was done and showed double inlet left ventricle, left transposition of the great arteries, preserved left ventricle systolic function, mild left atrioventricular valve regurgitation, PAB in position, PA gradient of 60mmHg, and no intracardiac clots or pericardial effusion. Patient was advised to terminate pregnancy after administration of dexamethasone. In two days patient developed severe reduction of diastolic blood flow. Emergency CS was done. During the CS patient was stable and the operation was smooth with no complications. Outcome was 1.43 Kg’s boy with APGAR score 6, 7, 8 at 1, 5, and 10 minutes. Baby was admitted to NICU for observation and kept there for 2 weeks and discharged in good health. Patient was observed for four days post-operative. She was discharged in good condition on oral medications, Metoprolol, Frusemide, and Clexan.

Discussion

The number of women reaching childbearing age after PAB procedure is steadily increasing [1, 3, 5]. This case is unusual because on literature review there is no case reports of any pregnancy in patient with PAB procedure. Accordingly, this is one of the first ever reports of successful pregnancy in such patients.

A double inlet left ventricle (DILV) or “single ventricle”, is a congenital heart defect appearing in 5 in 100,000 newborns, where both the left atrium and the right atrium feed into the left ventricle (Figure 1). The right ventricle is hypoplastastic or does not exist. Both atria communicate with the ventricle by a single atrio-ventricular valve. There is a big shunt left-right with a quickly evolutive pulmonary hypertension [1, 2]. Without life-prolonging interventions, the condition is fatal, but with intervention, the child may survive. Mortality is very high in the first 2 years, 85%, but after it decreases and between 2 and 15 years old the mortality is only around 9%. Few reach middle age [1-3]. Introduced in 1951, PAB procedure is still used in developing and under developed countries to treat DILV (Figure 2). Most patients who have not had the Fontan Operation (definitive treatment) will begin to show symptoms of cyanosis (external blueness caused by oxygen-poor arterial blood), fatigue, arrhythmias, and/or exercise intolerance, generally because of insufficient blood flow to the lungs through the pulmonary artery. They will also have a heart murmur because of pulmonary stenosis (narrowing of the outflow tract through which blood flows from the heart to the lungs) and/or because of atrioventricular valve dysfunction (the valve that connects the functioning ventricle with an atrium) [1, 2, 5, 6].

Our patient was unusual in remaining healthy during her early childhood without any corrective procedure. After the PAB procedure she was in good condition. On the other hand, the physiological changes of pregnancy influence the PAB circulation. The increase in blood volume and cardiac output, and the decrease in systemic vascular resistance together with the stress of pregnancy, represent a severe burden for the single ventricle [1, 2].

Unfortunately, our patient developed all possible complications of pregnancy on such delicate procedure and on top she developed severe reduction of diastolic blood flow. This, compelled us to proceed to emergency CS. Regional anaesthesia is considered the preferred anaesthetic technique for a patient with the PAB circulation because it is the least likely to interfere with pulmonary vascular resistance and ventricular function [1-3]. Removing the burden of pregnancy and its changes lead to, a welcomed, positive return in the patient condition to pre-pregnancy state.

Conclusion

PAB procedure is still used in developing and under developed countries to treat DILV. Physiological changes in pregnancy may potentially threaten the health of both the mother with PAB and her offspring, but routine tendency to discourage pregnancy was abandoned. With this case report, we can conclude that it is possible for those patients to carry on pregnancy but with vigilant and close follow up.

References

1. Bhimji, S. and J. Kupferschmid Pulmonary Artery
Illustrations

Illustration 1

Figure 1: DILV compared to normal heart

![Diagram of normal heart and double inlet left ventricle](image1)

Illustration 2

Figure 2: DILV with PAB

![Diagram of double inlet left ventricle with pulmonary artery banding](image2)