



## PULMONARY VALVE STENOSIS

### Background

Pulmonary stenosis (PS) may be valvular, supra- or subvalvular. The most common form of PS is valvular which is congenital in origin and usually occurs as an isolated sporadic defect. Valvular PS, however, can occur in the setting of an autosomal dominant syndrome such as Williams, Noonan or Alagille Syndromes, as part of tetralogy of Fallot or following maternal infection with Rubella.

The pulmonary valve in valvular PS is typically thin and pliable and usually domes in systole due to varying degrees of fusion of the commissures. Less commonly, the pulmonary valve may be thickened and dysplastic and exhibit decreased mobility of its cusps without commissural fusion. As part of a tetralogy of Fallot repair or severe valvular PS, patients may have undergone a pulmonary valvotomy leaving them with severe free pulmonary regurgitation and the sequelae of right ventricular (RV) volume overload.

Supra- or subvalvular PS usually occurs with one of the aforementioned genetic syndromes or can be associated with tetralogy of Fallot or a ventricular septal defect (VSD). Branch pulmonary arteries may also be involved.

Subvalvular PS involves narrowing of the infundibulum and is typically seen with other cardiac defects such as tetralogy of Fallot or a VSD. Double-chambered right ventricle is considered to be another form of subvalvular PS.

The severity of PS or right ventricular outflow tract (RVOT) obstruction is graded by the peak gradient across the RVOT: trivial <25 mmHg, mild 25-49 mmHg, moderate 50-79 mmHg and severe >80 mmHg.

Symptoms rarely occur in the absence of severe PS. These symptoms include: exercise intolerance, fatigue, dyspnea, presyncope or syncope, symptoms of right-sided congestive heart failure, chest pain from RV ischemia or symptoms related to arrhythmias.

Trivial and mild valvular PS rarely progresses significantly. Symptomatic patients with severe (and sometimes moderate) PS will often require a percutaneous balloon (or surgical) valvotomy. Both supra- or subvalvular PS have been reported to progress and should be followed closely long-term as they may require an intervention should they become moderate-to-severe or the patient develops symptoms.

### Effects of pregnancy related hemodynamic changes

Pregnancy is associated with significant hemodynamic changes including increases in blood volume and cardiac output. (see Cardiovascular Changes During Pregnancy) The hemodynamic effects of pregnancy are usually well-tolerated in patients with PS. If the PS is severe, the increased volume load of pregnancy on a stiff and hypertrophied pressure-loaded RV may predispose to right-sided heart failure, significant tricuspid regurgitation and arrhythmias particularly if there is underlying RV dysfunction.

## Maternal Cardiac Complications

In general, most women with PS, even severe PS, usually tolerate pregnancy well without cardiac complications. (1,2) Other cardiac characteristics can have an impact on outcomes (see General Considerations).

There is evidence to suggest that this population may be at increased risk for non-cardiac and neonatal complications based on one published report. In the largest reported series of 108 pregnancies in 51 patients with isolated valvular PS, a high number of complications were observed including pregnancy-induced hypertension, pre-eclampsia, eclampsia, miscarriages, thromboembolic complications and premature rupture of membranes. (3)

## Fetal Complications

Favorable fetal and neonatal outcomes have been reported in asymptomatic or mildly symptomatic patients with isolated PS of varying degrees of severity and normal RV function. (2) Conversely, a very high rate of non-cardiac complications (prematurity and cardiac defects) and mortality were reported in offspring of women with uncorrected isolated valvular PS. (3)

## Management Strategies

### Preconception counselling/Contraceptive methods

Women should be advised to meet with their cardiologist prior to becoming pregnant so that their baseline status can be determined. Ideally, symptoms related to PS or moderate or severe PS with associated RV dysfunction should be addressed prior to pregnancy with a percutaneous or surgical intervention, if necessary.

The indications for surgical or interventional procedures prior to pregnancy should follow recommendations in current guidelines. (4,5,6)

Asymptomatic women with isolated PS and normal RV function are likely to tolerate pregnancy well without incident. (1,2) It may be useful to perform a treadmill or cardiopulmonary exercise test as a screening test for women with underlying RV dysfunction or minimally symptomatic PS to help assess how they will tolerate the hemodynamic stress of pregnancy (3).

A discussion about contraceptive options is appropriate in women with PS. In general, most forms of contraception are safe in women with PS. (see Contraception)

Transmission of congenital heart disease to offspring should be discussed. The risk of transmission of congenital heart disease is approximately 5% for women with isolated PS, compared to a background risk of approximately 1% in the general population. For women with Williams, Noonan and Alagille syndromes, transmission to offspring differs. These syndromes have autosomal dominant transmission and confer up to a 50% chance of transmission to the fetus.

Medication use should be reviewed if a woman is contemplating pregnancy or is pregnant. The MOTHERISK website (<http://www.motherisk.org>) is an excellent resource.

## **Ante-partum care**

Women with symptoms, RV dysfunction or PS that is moderate or greater in severity should be followed at a hospital center that specializes in high-risk pregnancies. Coordinated care between a heart specialist, a high-risk obstetrician and an anesthetist should be implemented. The frequency of follow-up visits and echocardiograms should be dictated by the specialists.

In women with low risk features, depending on their preferences, antenatal care and delivery can be performed at non-specialized centers.

Although a percutaneous balloon valvotomy is preferable to perform prior to conception, when necessary, percutaneous balloon valvotomies with uterine shielding during pregnancy have been performed. (7) A surgical intervention is usually required to manage subvalvular PS and some forms of supra-valvular PS.

All women with PS should be offered fetal echocardiography at 20 weeks gestation.

## **Labour and delivery**

For women with moderate or severe PS or RV systolic dysfunction, labor and delivery should be planned carefully with a multidisciplinary team well in advance. It is important to communicate the delivery plan to the woman and to other physicians involved in her care. The best delivery plan is not useful if information is not readily available when needed.

Generally, vaginal delivery is recommended. Good pain management for labour and delivery is important in order to minimize maternal cardiac stress.

For most deliveries of women with PS, maternal monitoring is not required.

In general, endocarditis prophylaxis at the time of labour and delivery is not recommended in women with PS. However, some experts continue to administer antibiotics because they feel that the risks of adverse reactions to antibiotics are small and the risk of developing endocarditis has major health consequences.

## **Post-partum care**

The hemodynamic changes of pregnancy may take up to six months to normalize. Women should be seen early after pregnancy (usually within 6-8 weeks). The frequency of additional follow up visits should be dictated by the clinical status of the women.

## References

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