Valvular pulmonic stenosis. Diagnosis and therapy reviewed.

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Summary

In this thesis the diagnosis and therapy of Valvular Pulmonic Stenosis (VPS) are reviewed. One reason for this is founded in the improvement in diagnostic techniques that has taken place in the past decades. Another reason is the change in possible therapy due to the development of Percutaneous transluminal Balloon Valvuloplasty (PBV).

In chapter 1 a short introduction is given of the place of congenital VPS in the group of known congenital cardiac malformations. This place is marked by a longstanding experience in noninvasive diagnostic techniques. However, the more recent development of two-dimensional echocardiography outweighs the value of older noninvasive techniques, although not yet demonstrated in adult patients with VPS. The development of balloon dilating therapies for narrowed vessels and narrow structures in and around the heart altered the therapeutic scope. Until recently surgery was the treatment of choice with rather circumscript criteria for admission. The relative ease of a valvuloplasty procedure raises the question if the admission criteria should be lowered.

In chapter 2 an extensive overview of the diagnosis of VPS is given.

It begins with a short introduction of the embryology and pathophysiology of the pulmonary valve. Two types of stenotic valves can be distinguished: the dome shaped valve with fusion of the valve leaflets and the dysplastic valve with grosly deformed and thickened valve leaflets. The last type is frequently combined with narrowing of the valvular ring. A special clinical syndrome is the critical VPS of the newborn characterized by severe right to left shunting on atrial level and diminutive right ventricle.

The use of the stethoscope plays a key role in detecting VPS because most patients don't have spontaneous complaints. The hallmarks during physical examination are the ejection murmur over the left upper sternal border and the widened splitting of the second heart sound with diminished pulmonary component. The ECG is mostly normal, only in patients with severe VPS right ventricular hypertrophy is the dominant finding. Chest X-rays often show an enlarged pulmonary artery segment but are otherwise normal. Phonocardiograms are often used to offer a visible display of sounds and murmurs. Measurements of distances between heart sounds, for instance the interval of the aortic and pulmonic component of the second heart sound, are used to estimate the severity of the valvular stenosis. Echocardiograms are used to visualize the pulmonic valve and right ventricular anatomy. Doppler echocardiography can measure flow velocity in the pulmonary artery and permits rather precise computation of transvalvular pressure differences. Catheterization of the heart is of course the method that can establish with certainty the diagnosis

of VPS and its severity, but it is at the same time the most cumbersome procedure for the patient. Distinction from other or accompanying congenital anomalies like atrial septal defect can reliably be made with this technique.

After making the diagnosis of VPS the next step is to assess the severity of the valvular stenosis. The ECG is not very reliable to predict the peak systolic right ventricular pressure. A combination of clinical variables with ECG items improves the relationship with invasive measurements. The VCG is more precise in quantitating right sided ventricular forces but it still has too little specificity. The measurement of the 2A-2P interval in the phonocardiogram cannot give the clue about severity of the stenosis because in pure infundibular stenosis the second heart sound is also split widely and not related to transvalvular pressure difference. In conclusion the older noninvasive techniques make the obsever aware of the possibility of VPS but are not able to assess with certainty its severity.

The development of doppler echocardiography enables the measurement of flow velocity in the pulmonary artery. With the help of the Bernoulli equation calculation of transvalvular pressure difference is possible.

In children with VPS there have been published promising reports about the good relation between the calculated echo doppler pressure differences and the invasive measured ones. In comparison with the older noninvasive techniques the doppler echocardiogram is superior in assessing the severity of VPS.

To differentiate pure infundibular stenosis from VPS or to assess the severity of associated infundibular hypertrophic narrowing in VPS the echocardiogram can be helpful. Only if this technique produces inconclusive results invasive investigations are necessary.

There are many data lacking concerning the long term follow-up in adults with VPS. For children with VPS it is known that progression of the valvular stenosis is not the rule.

In chapter 3 the different forms of therapy are discussed. The surgical repair by transventricular valvotomy according to Brock was reported in 1948. Later on the technique of inflow occlusion of the right heart with transpulmonic approach was developed. The use of the cardio-pulmonary bypass enabled the surgeon to resect the hypertrofied infundibulum if necessary. The dome shaped stenosis is treated by valvotomy, the dysplastic valve is partly resected and sometimes an annuloplasty is necessary. The overall mortality concluded from several reports is about 4% but there exists a negative influence on this figure by the mortality of the newborn with critical VPS. Also the improvement in surgical and anesthesiological techniques and materials result in a much lower mortality figure in the more recent reports. After surgical valvotomy a transvalvular pressure difference of 20-30 mmHg is considered as a satisfactory result.

Percutaneous transluminal Balloon Valvulopasty (PBV) as treatment for VPS was introduced by Kan. Some resemblance is present with the surgical technique

according to Brock: the valve is split without direct vision. The advantage of a procedure with a balloon catheter is evident in comparison with a surgical procedure, even more clear if the low complication rate is considered. The first results are promising, but the results on the long-term are not known yet. The transvalvular pressure difference after PBV is the same magnitude as after surgery.

Until so far the same criteria are used for surgery and PBV: transvalvular pressure difference of more than 50 mmHg, pulmonic valve area smaller than 0.7 cm^2 or right ventricular peak systolic pressure more than two-third of left ventricular peak systolic pressure. Because of the low complication rate there is a trend to lower the admission criteria for PBV compared to surgical treatment.

After this survey of the literature on diagnosis and therapy two chapters with own research material are presented. In chapter 4 a study of the natural history of adult patients with VPS is described with emphasis on the noninvasive diagnostic techniques. A number of 34 patients with mild VPS were willing to come to the outpatient clinic for noninvasive follow-up investigations. Of this group 24 patients gave permission for invasive reevaluation and the diagnosis of VPS only was accepted if at least one invasive exemination either at the start or at follow-up was present. The mean clinical follow-up time was 20.4 years, mean starting age was 19.2 years. The patients had no spontaneous complaints during the follow-up period.

Auscultatory data showed a sligt decrease in loudness of the ejection murmur. In 30 0f 34 patients the ECG stayed normal, in two patients signs of right ventricular hypertrophy disappeared. Chest X-rays showed in 61% prominence of the main pulmonary artery, a frequency somewhat lower than found in the literature. From the phonocardiograms the 2A-2P interval, the Q-click time and the Q-peak time of the ejection murmur were correlated with the invasive transvalvular pressure difference. The correlation coefficients were disappointing and low. No changes in phonocardiographic variables were seen during follow-up. Also the invasive data did not show significant shifts, even when only paired data were considered.

The echocardiogram is the most promising noninvasive investigation in VPS. There were no follow-up data available because at the starting point of the study no echocardiography was available. Adequate M-mode recordings of the pulmonary valve were obtained in 97% of the patients. In only 15% of them an abnormal pulmonary valve was detected, maybe because only patients with mild VPS were studied. In 10 patients doppler measurements of the maximal flow velocity in the pulmonary artery were obtained. The correlation coefficient of the calculated transvalvular pressure differences and the invasive measured ones was 0.79.

The conclusion from this study is that no progression of the stenosis in VPS can be detected in adult patients with mild VPS. The abnormal pressure load of the right ventricle is endured without clinical signs and symptoms.

In chapter 5 the experience of PBV for VPS of the University Hospital of Groningen is described. From august 1984 38 patient were treated with PBV, 30 children and 8 adults. No serious complications were noticed.

The mean transvalvular pressure difference decreased from 51 mmHg before PBV to 21 mmHg after PBV, functional valve area increased from 0.53 to 0.94 cm2. One patient was redilated because a too small balloon was used in the first attempt.

From this group 17 patients underwent invasive evaluation after about one year. The functional valve area further increased from 0.97 to 1.29 cm^2 but this was not significant. This further tendency to increase is probably caused by a decrease in secondary infundibular hypertrophy. No substantial decrease in valve area was seen.

The main auscultatotory finding after PBV was a decreased intensity of the ejection murmur. A significant decrease in right sided electrical forces on the ECG was seen at a period of three months follow-up. One year after PBV no further significant changes in the ECG occurred.

In 94% of the patients abnormalities of the pulmonary valves were detected with the two-dimensional echocardiogram. Probably the fact that in this group only patients with moderate to severe VPS were gathered can explain the difference in observations of the group with mild VPS in chapter 4.

The doppler echocardiogram provided calculations of the transvalvular pressure differences which correlated very well with the invasive measurements. When all the measurements before and after PBV were taken together a correlation coefficient of 0.88 was found, although the echo measurements and the invasive ones were not recorded at the same time.

The incidence of pulmonary valve incompetence before and after PBV increased from 12% to 48%, measured with doppler echo. With auscultation this rise was much less impressive. Although in our patient material this pulmonary insufficiency was judged as nonsignificant long term follow-up is needed to verify this judgement.

The question if restenosis will occur on the long term has to be answered in the future. In comparison with the surgical technique according to Brock it is expected that the improvements with PBV will have a permanent character.

In the epilogue (chapter 6) the results of chapter 4 and 5 are summarized and discussed in a more general setting. The main change in the diagnosis of VPS is caused by the use of doppler echocardiography. Only this instrument can assess the severity of VPS with sufficient reliability. Its use in differentiating from other congenital anomalies is invaluable.

Because of the good results with PBV in patients with VPS this treatment has become the treatment of choice with exception for the newborn with critical VPS. The relative ease and the low complicaton rate of the PBV procedure raises the question if also patients with mild VPS should undergo this treatment.

Support to this point of view is found in haemodynamic arguments. The right ventricular pressure rise during exercise to systemic level in patients with mild VPS can be the cause of myocardial fibrosis on the long term. On the other hand the natural history of adult patients and children with mild VPS is very reassuring. Moreover, the induction of pulmonary insufficiency after PBV can have negative influence on right ventricular function on the long term. Determination of right ventricular function under different conditions will possibly result in more reasonable admission criteria for PBV in the group of patients with mild VPS.