Guideline Pulmonary Valve Stenosis

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1. Definition, Classification, Basic information

Distinction between

- **Critical pulmonary stenosis of the newborn**

Severe stenosis with duct dependant pulmonary circulation, cyanosis, decompensation of the right ventricle

- **Typical pulmonary valvar stenosis during childhood**

Fusion of the commissures, pulmonary valve annulus usually of normal diameter, poststenotic dilatation of the main pulmonary artery

- **Dysplastic pulmonary valve**

Myxomatous thickend valve with 3 hypomobile cusps, pulmonary valve annulus usually hypoplastic, no or only mild poststenotic PA dilatation, often supravalvar stenotic component, main PA and PA branches of normal size or hypoplastic

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2. Epidemilology, prevalence, associated leasions, syndromes

- Isolated pulmonary valve stenosis acounts for ~ 6% of CHD (1)
- In 10-15% dysplastic valves
- Association with various other CHD, in particular ASD, peripheral PS
- Assiciation with various genetic and chromosomal syndromes; most frequently
  - Noonan (dysplastic PV)
  - Williams-Beuren
  - Allagille

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3. Pathophysiology and hemodynamics

- Hemodynamically RVOTO resulting in elevated RV pressure (up to suprasystemic)
- Compensatory RV hypertrophy
- With severe stenosis inability to adequately increase CI under exercise
- Most severe forms with coronary hypoperfusion in subendocardial myocardium leading to fibrosis and RV myocardial insufficiency and dilatation
- Increased RV enddiastolic pressure and reduced RV compliance (hypertrophy) result in elevated RA pressures and subsequnetly RA dilatation
- In critical PS right-left shunting via PFO with central cyanosis

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4. Physical examination and leading symptomes

In **critical PS** ductal closure leads to severely reduced pulmonary perfusion with life-threatening hypoxia.
Clinical exam: central cyanosis, signs of congestive heart failure, diminished ejection murmur (decompensated RV)

In **typical PS of childhood**
Clinical exam: systolic ejection type murmur with p.m. in 2nd left intercostal space, ejection click, 2nd heart sound widely split, pulmonary component scarcely audible, palpable thrill

With **dysplastic valves**
No ejection click

Possible symptoms of severe un-treated PS
(esp. with exercise) dyspnea, exercise intolerance, fatigue, (mild) cyanosis, hepatosplenomegaly, pronounced jugular venous pulsation, chest pain, palpitations or syncope

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5. Diagnostic investigation
Aim: to establish the diagnosis and degree of severety, to exclude or varify additional anomalies, like sub- or supravalvar stenosis or pheripheral PS

Echocardiography:
Fetal echocardiography

- Serial follow-up (progression?)
- Also with flow velocity of 2-3 m/s significant gradient with need for intervention after birth possible (reduction in pulmonary vascular resistence!)
- Assess PV anatomy and flow, PA flow, RV size and hypertrophy, TV insufficiency
- Prenatal balloon valvuloplasty to prevent RV hypoplasia? (relation of TV to MV and RV to LV)
- Prenatal counceling must include possible syndromatic association (e.g. inadequat hypertrophy with possible additional cardiomyopathy/ Noonan)

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Checklist prenatal echocardiography		
	Mild to moderate fetal PS	Severe fetal PS
Pulmonary valve	Valve thickend, dysplastic, variable poststenotic dilatation	
Doppler	Aliasing, vmax < 2m/s	Aliasing, vmax > 2m/s, dependent on RV function, or retrograde color Doppler via duct
RV	normal	Myocardial hypertrophy, reduced RV volume/ hypoplasia
TV		TR up to grade IV

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Postnatal echocardiography

- PV: diameter and morphology, systolic Doppler RV-PA gradient (attention: degree of stenosis may be underestimated in pulmonary hypertension of the newborn!), PV insufficiency?
- PA: diameter of central PA, poststenotic dilatation?, additional supravalvar or peripheral PS?
- TV: diameter, estimate RV pressure with tricuspid insufficiency
- RV: size, morphology (bi- or tripartite) and function, RVOT with (reactive) subvalvar obstruction
- Atrial septum: shunt?, if possitive: shunt volume and direction
- Additional anomalies?, relative PS (e.g. ASD, AV-malformation)

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Echocardiographic assessment of severety and therapeutic consequence		
Degree of severety	Max. systolic Doppler gradient (at rest)	Therapeutic consequence
Mild	< 40 mm Hg	No treatment indication
Moderate	40-60 mm Hg	Treatment may be initiated
Severe	> 60 mm Hg	Treatment should be initiated

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5. Diagnostic investigation

- Catheterisation: for therapy (see below)
- Cardiac MRI: usually not necessary, individually based with poor echo windows and suspicion of additional lesions (e.g. peripheral PS)
- Cardiac CT: usually not necessary
- Chest X-ray: usually not necessary
- ECG: signs of RV hypertrophy usually correlating with RV pressure, possibly signs of RA hypertrophy and right axis deviation
- Pulse oximetry: documentation and degree of cyanosis
- Differential diagnosis: tetralogy of fallot, pulmonary atresia with intact ventricular septum, PPAH of the newborn, absent antegrade flow with normal PV (“functional atresia”)

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6. Therapy

Indication for treatment

- Always in critical PS (irrespective of gradient)

- In older children: always with symptoms; in asymptomatic patients treatment (balloon valvuloplasty) can be offered with max. systolic Doppler gradient > 40 mm Hg, with severe PS (max. systolic Doppler gradient > 60 mm Hg) treatment should be initiated

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6. Therapy

Medical treatment

- Significant congestive heart failure should be treated with apropriate medication (e.g. catecholamines, phosphodiesterase inhibitors, if necessary diuretics) without delaying a pulmonary valve intervention

- In critical PS intravenous prostaglandine infusion should be initiated to re-open the duct or to keep it open until the planed intervention (in prenatally detected cases start prostaglandine right after birth)

- In critical PS irrespective of type of PV intervention (catheter or surgery) in up to 20% an alternative source of pulmonary blood supply is temporarily needed, also after successful intervention. This can be due to the restrictive RV physiology (prostaglandine infusion, ductal stenting, aorto-pulmonary shunt)

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6. Therapy

Treatment by catether intervention (balloon valvuloplasty)

- Method of choice for treatment of isolated PS without dysplastic valve
- The choosen balloon diameter relates to the valve annulus and should usually be choosed around 130% (120-150%)
- Dysplastic valves usually respond less or not at all to balloon vavluloplasty due to the underlaying pathology

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6. Therapy

Surgical treatment

- Indicated if catheter interventions fails or as primary treatment with dysplastic valves

- Aim: not only elimination of valvar stenosis but also maintainance of valvar competence (importance for long-term follow-up!)

- Under extracorporal circulation: open commissurotomy of fused cusps, mobilisation of thickend cusps by valve shaving or partial excision, if necessary enlargement of valve annulus and concomittant myectomy with or without patch plasty of the RVOT with (reactive) subvalvar stenosis

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6. Therapy

Risk of intervention and treatment results

- Generally **balloon valvuloplasty of PS** is an effective treatment with good long-term prognosis

- Freedom of re-intervention (more often for re-stenosis as for insufficiency) is 95, 88 and 84% after 5, 10 and 20 years. Re-Intervention rate is higher with dysplastic valves

- Mortality with BVP is low, also in neonates (0-0.5%). Serious vascular access problems are rare.

- Freedom of re-intervention (more often for insufficiency as for re-stenosis) after **valve surgery** is 98, 94, 88, 71 and 56% after 5, 10, 20, 30 and 40 years

- Operative mortality is given at 0-1% in infants and older children and higher in neonates

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7. Follow-up

Clinical follow-up

- Beside immediate residual stenosis in 5-10% recurrent stenosis (mostly within the 1st year)
- In 10-40% pulmonary insufficiency, initially usually insignificant, but may progress over time (esp. after puberty)
- Clinical follow-up with echo, (Holter) ECG, spiroergometry and MRI (quantification of RV volume, function and pulmonary regurgitant fraction) also beyond childhood age
- Endocarditis prophylaxisis not indicated

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7. Follow-up

Sports and exercise

- In asymptomatic patients with peak Doppler gradient < 40 mm Hg competitive sport possible; should be reassured and encouraged for normal physical activity

- Patients with peak Doppler gradient > 40 mm Hg should avoid competive sports before interventional therapy

- In patients with significant pulmonary regurgitation and RV dilatation sportive activity with low intensity should be recommended (see guideline on pulmonary valve insufficiency)

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7. Follow-up

Gown-ups with congenital heart disease (GUCH)

- Same treatment indications apply for adolescents and grow-ups

- Long-term follow-up of (residual) pulmonary stenosis and regurgitation with acessemnt of RV volume and function (MRI).

- Tricuspid insufficiency and occur or progress

- Depending on RV pressure and volume load supraventricular and ventricular arhythmia may develop

- In women with PS without right-left shunting (at atrial level) there is no increaed risk with oral contraception. With right-left shunting a mare gestagen formulation or non-hormonal form of contraception should be used due to the thrombo-embolic risk

- In mild pulmonary stenosis (without treatment indication) occupational choice is generally not restricted

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7. Follow-up

Pregnancy

- Asymptomatic women with peak Doppler gradient < 40 mm Hg have no increased (maternal) risk during pregnancy

- Close follow-up is necessary since gradient can increase during 2nd and 3rd trimester of prgnancy due to increased circulatory volume.

- Vaginal delivery is generally well tollerated and should be strived for

- In women with a peak Doppler gradient > 40 mm Hg an (re-)intervention should be carried out before planned pregnancy

- Given the indication, a balloon valvuloplasty can be done effectively in pregnant women with lead protection of the womb or under mare echo guidance

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