Guideline
Peripheral Pulmonary Artery Stenosis

H. Bertram, MHH, Hannover
M. Schneider, DKHZ, St. Augustin
A. Horke, MHH, Hannover
DGPK guideline committee
Peripheral Pulmonary Artery Stenosis

**Definition**

- Stenosis of the pulmonary artery *distal to the pulmonary trunk*

Broad morphologic spectrum:
- isolated / part of CHD
- single / multiple
- congenital / iatrogenic
- elastic / scar forming
- circumscript / diffuse hypoplastic
Peripheral Pulmonary Artery Stenosis

Epidemiology

Underlying disease:

- Williams-Beuren-syndrome
- Alagille syndrome
- Noonan syndrome
- congenital rubella syndrome
- systemic vasculitis (Takayasu, Behcet)
- Ehlers-Danlos syndrome
- Silver-Russel-syndrome
Peripheral Pulmonary Artery Stenosis

**Epidemiology**

**Part of Congenital Heart Disease (CHD):**
- TOF / DORV / PA-VSD, multifocal esp. with hypoplasia / 'coarctation' of proximal branch pulmonary arteries

**Iatrogenic pulmonary artery stenosis:**
- s/p palliative surgery
  - modif. BT-shunt
  - PAB
  - CPA
- s/p surgical repair of CHD using patch augmentation of central PAs or RV-PA-conduits

s/p surgical repair of DORV/TOF, with 'coarctation' of central PA
Peripheral Pulmonary Artery Stenosis

**Pathophysiology - Hemodynamics**

- **Pressure load of the subpulmonary ventricle:** its extent depending on number and severity of stenoses
- Myocardial hypertrophy
- Subsequent pulmonary valve and/or tricuspid valve regurgitation
- Underperfusion of the poststenotic vascular bed (=> collaterals!)

Later stages (> moderate / severe disease):
- Ventricular dilatation
- Right atrial enlargement due to RV diastolic dysfunction
- Arrhythmias
- Right heart failure with hepatomegaly and ascites
- Loss of lung parenchyma due to totally occluded segmental PAs
Peripheral Pulmonary Artery Stenosis

Clinical features

Clinical presentation
- **systolic murmur** conducted into the respective lung
- **peripheral bruits** over the back or on either lateral side of the chest
- 'free of symptoms' to right heart failure

Advanced disease:
- parasternal heave / pronounced precordial pulsation due to RV pressure load and dilation
- **Atrial (tachy-)arrhythmias** due to secondary RA enlargement (diastolic RV dysfunction)
- **Cyanosis** secondary to intracardial right-to-left-shunts
- Hepatomegaly, ascites, peripheral edema

Differential diagnoses
- Peripheral Pulmonary Artery Stenosis as part of CHD (TOF / PA-VSD / DORV)
- Subvalvar, valvar, supravalvar pulmonic stenosis
- Primary Pulmonary Arterial Hypertension
- Adults: chronic pulmonary thrombembolic disease
Peripheral Pulmonary Artery Stenosis

Diagnostic work-up

**Aim**

⇒ Confirmation of the diagnosis
⇒ Displaying the entire pulmonary arterial system to define the amount and severity of the lesions

**Echocardiography:**

- Confirmation of RV pressure load, pulmonary and / or tricuspid valve regurgitation, intracardial shunts due to septal defects
- Exclusion of congenital heart disease / RVOTO (valvar, sub-/supravalvar)
- Morphologic description of stenoses close to the pulmonary bifurcation ('central'); grading the severity using Doppler flow pattern

**ECG:** (advanced disease)

- Right ventricular myocardial hypertrophy / dilation
- (incomplete) Right Bundle Branch Block
- Right atrial enlargement; atrial tachyarrhythmias
Peripheral Pulmonary Artery Stenosis

Diagnostic work-up

**Chest X-ray:** (advanced disease)
- Cardiomegaly / prominent pulmonary trunk
- (may detect) relative flow discrepancy between the two lungs

**Magnetic resonance imaging:**
- MR angiography is superior to echocardiography for imaging multiple peripheral pulmonary artery stenoses
- Demonstration and quantification of secondary right ventricular and right atrial enlargement, pulmonary valve or tricuspid valve regurgitation
- Quantification of relative flow discrepancy between right and left lung
- Drawbacks: - (long) sedation / general anaesthesia in infants / young children
  - multiple artefacts in postoperative (→ valves) / postinterventional (→ stents) pts.
Peripheral Pulmonary Artery Stenosis

Diagnostic work-up

**Computed tomography:**
- high resolution CT angiography is superior to echocardiography and MRI for imaging of multiple peripheral pulmonary artery stenoses
- increasingly used for interdisciplinary management discussions of complex cases
- short acquisition times enabling CT diagnostic without long sedation / anaesthesia
- Drawbacks: - radiation exposure
Peripheral Pulmonary Artery Stenosis

Diagnostic work-up

Cardiac catheterization:

- Angiography is **still gold-standard** for detailed imaging; (super-)selective injection delineate the exact anatomy, esp. in complex lesions of lobar / (sub-)segmental level
- Balloon sizing may provide additional accuracy in terms of severity and extension of the stenotic segment(s)
- Interventional therapy can be performed during the same session
- Drawbacks: - invasive - radiation exposure
Peripheral Pulmonary Artery Stenosis

Diagnostic work-up: Catheterization

Selective angiography and balloon interrogation in complex bilateral pulmonary artery stenoses
## Peripheral Pulmonary Artery Stenosis Management

### Indications for treatment

- **Reduction of vessel diameter** ≥ 40% or reduction of vessel diameter ≥ 30% in case of additional RV volume load (ASD; tricuspid regurgitation; pulmonary regurgitation)
- In patients after cavo-pulmonary anastomoses (s/p Glenn or Fontan) a PA stenosis should be considered for therapy even if less than 30% diameter reduction
- Bilateral PA-stenoses resulting in RV pressure load of > ½ systemic
- Significant stenosis with measurable gradient of > 20 (30) mm Hg
- Branch-PA-stenosis resulting in the reduction of ipsilateral lung perfusion < 20 (-30)%

### Aim

- Relief of PA stenosis
- Reduction in RV pressure load to < ½ systemic
- Improvement of ipsilateral lung perfusion (to > 35 %)
- Improvement of peripheral lung perfusion to reduce hemodynamic effects of pulmonary valve regurgitation
Peripheral Pulmonary Artery Stenosis
Management

**Drug treatment:**
- **there is no medical treatment for relief of peripheral pulmonary artery stenosis**
- secondary right heart failure should be treated according to guidelines
- Platelet aggregation inhibitors should be considered after surgical or catheter intervention

**Williams-Beuren syndrome:**
- Stenoses might recede over time
  => 'wait and watch' strategy might be considered if RV pressure load is acceptable
Peripheral Pulmonary Artery Stenosis Management

**Surgery**
- Bifurcation
- Branch PA

**Catheter Intervention**
- Branch PA
- Lobar PA
- Segmental PA
- Bifurcation

An interdisciplinary discussion [surgeon and interventionalist with training and expertise in CHD] based on the patient’s individual anatomy and history is strongly advised!
Peripheral Pulmonary Artery Stenosis
Management - Surgery

- reconstruction of central PA using **patches**
- predominantly performed in combination with surgical repair of CHD or insertion of RV-PA conduits
• in patients with severe hypoplasia of the pulmonary artery tree, insertion of an aorto-pulmonary shunt may promote vessel growth
Peripheral Pulmonary Artery Stenosis
Management - Surgery

Surgical Techniques for Repair of Peripheral Pulmonary Artery Stenosis

no standard therapy !

30 d mortality for branch PA reconstructuion (plasty) central / peripheral: 4 – 6 %
https://echsacongenitaldb.org/gs_report /  EACTS / STS database
Balloon Angioplasty (PTA)

- applicable for central PA stenoses as well as lobar and segmental stenoses
- effective in postoperative scar formation
- indicated particularly in very small patients or those with complicated anatomy, in whom primary stent implantation is not a viable option
- high pressure balloons (> 10 [up to 40 !] atm) and 'cutting balloons' may be considered in lesions refractory to standard PTA
Peripheral Pulmonary Artery Stenosis
Management – Catheter Intervention

Balloon Angioplasty - outcome

- balloon dilation alone (standard PTA) may produce complete resolution of the obstruction / normalization of flow to the poststenotic area in ~ 30-40 % of lesions
- high pressure balloons, cutting balloons and stents may increase success rates up to 80 %
- the improvement achieved (50 % improvement in diameter / 50 % decrease in gradient) is often transient
- best results in (postoperative) central stenoses
- worst results in young children with multiple bilateral peripheral lesions
- reinterventions are frequent
Peripheral Pulmonary Artery Stenosis
Management: Ballon angioplasty - case

middle lobe artery
upper lobe artery
small vessel tear
Peripheral Pulmonary Artery Stenosis Management – Catheter Intervention

Risks and Complications

Multiple high pressure balloon angioplasties of lower lobe arteries and middle lobe artery in a 2 year-old boy with complex bilateral pulmonary artery stenoses and suprasystemic RV pressure load

Reperfusion injury

@ 1 d

@ 3 d

@ 6 d
Peripheral Pulmonary Artery Stenosis
Management – Catheter Intervention

Stent placement

- Treatment of choice, when
  - balloon dilation alone is inadequate
  - the stent used is capable of being dilated to
    the adult diameter of that vessel

- applicable for branch PA stenosis / lobar artery
  stenosis / segment artery stenosis

- improvement of stent design and interventional
  techniques enable stent angioplasty of
  bifurcational stenosis as well
Peripheral Pulmonary Artery Stenosis
Management – Catheter Intervention

- Small pulmonary artery stents which lack the potential to achieve adult size may be used in critically ill postoperative patients with hemodynamic compromise due to branch PA stenosis or whenever the surgical risk is considered too high [bailout]

- Permanent resolution of pulmonary artery stenoses may be achieved in ~ 80% of lesions; reinterventions might be necessary during f/u, depending on patient’s age and weight
US multi-institutional registry: Catheter interventions (balloon + stent) in branch PA stenosis → 10% incidence of high-level AE risk factors:
• hemodynamic vulnerability
• young age
• use of cutting balloons
• lower operator experience

Peripheral Pulmonary Artery Stenosis Management – Catheter Intervention

Risks and Complications

- Tearing a pulmonary artery (dissection, rupture, bronchial bleeding): Balloon (overdilation !) > Stent
- Wire perforation & Arrhythmia: Stent > Balloon (stiffer wires and delivery systems)
- Malpositioning of the stent => „jailing“; stent migration

- 30 d mortality 1-2% (balloon angioplasty and stent placement as well) (UK data 2013-2016, NICOR)
Peripheral Pulmonary Artery Stenosis
LPA Stent placement - case

Dg: TOF, right aortic arch, hypoplastic LPA, left duct
- Balloon Angioplasty @ 4 weeks
- left modif. BT-shunt @ 6 weeks
- Stent angioplasty LPA @ 8 weeks
- Surgical repair + stent explantation + LPA reconstruction @ 2 mo
- 2nd LPA stent @ 4 y
- Pulmonary valve replacement (Contegra), stent explant., tricuspid valve reconstruction, and 2nd LPA patch @ 10 y

- Diminutive prox. LPA on echo => recatheterization 8 months after last operation
Peripheral Pulmonary Artery Stenosis

LPA Stent placement - case

3rd Stent in LPA @ 11 y (10 mm balloon)

Stent redilation @ 15 y (15 mm)
Peripheral Pulmonary Artery Stenosis

PA stent placement in univentricular circulation - case

HLHS (AA, MS), s/p TCPC [intracardiac lateral tunnel] with fenestration, s/p PLE;

A: Angiography in intracardiac 'lateral tunnel' demonstrating diminished perfusion of LPA
B: Selective angiography in central PA displaying length and diameter of the stenosis
C: Placement of stent
D: Angiography in superior cavo-pulmonary anastomosis demonstrating improved LPA perfusion without vessel damage
Peripheral Pulmonary Artery Stenosis
Hypoplastic central PA - case

- PA/VSD, hypoplastic PAs, absent MPA, right AoArch, left duct, 'coarctatio pulmonalis'
- s/p modif. left BT-shunt
- Surgical repair @ 9 mo: ASD-/VSD-closure + 12 mm Contegra + bifurcation patch

- Postoperative persistent peripheral PS
- suprasystemic RV pressure
- rapid Contegra dilation
  => re cath @ 11 mo
Peripheral Pulmonary Artery Stenosis

Bifurcation stenosis after RV-PA-conduit - case

Stent in left PA 7 mm

PA-angiography after bilateral stent placement

Stent in right PA 6 mm

Stent adaptation with 8 mm balloons @ 26 mo
Peripheral Pulmonary Artery Stenosis
LPA rehabilitation in univentricular circulation - case

Mesocardia, PA/VSD, MGA, ASD, I-SVC
• s/p neonatal left BT-shunt
• s/p bilateral CPA @ 11 months => occlusion of I-SVC + central PA/LPA
• s/p TCPC 'on one lung' @ 8 years

- referred for Dx + Tx of asthma
- CT thorax, followed by re-cath @ 14 y
- Wire and subsequent catheter recanalization
- Multiple balloon angioplasties
Peripheral Pulmonary Artery Stenosis
LPA rehabilitation in univentricular circulation - case

2 stents in central PA
Re-angioplasty after 6 months

Re-cath @ 19 y
5 years after LPA rehabilitation
# Peripheral Pulmonary Artery Stenosis Management: Summary

<table>
<thead>
<tr>
<th></th>
<th>Balloon Dilation</th>
<th>Temporary Stent</th>
<th>Permanent Stent</th>
<th>Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age</strong></td>
<td>all age groups</td>
<td>infants</td>
<td>beyond infancy</td>
<td>all age groups</td>
</tr>
<tr>
<td><strong>Localization</strong></td>
<td>central &amp; peripheral</td>
<td>central &amp; peripheral</td>
<td>central &amp; peripheral</td>
<td>central</td>
</tr>
</tbody>
</table>
| **Advantages**   | - no permanent devices  
- standard PTA with low procedural risk  
- high efficacy in postoperative scar forming lesions | - rapid procedure with high efficacy and considerable low risk in patients with standard anatomy  
- postponing surgical (re-)intervention | - rapid procedure with high efficacy and considerable low risk in patients with standard anatomy  
- replacing (re-)operation  
- low risk of intimal proliferation in PA  
- larger stents might be placed during Hybrid Interventions | - PA reconstruction might be performed within surgical repair of CHD  
- larger stents might be placed during Hybrid Interventions |
| **Disadvantages**| - high rates of residual stenosis or restenosis  
- frequent reinterventions | - unavoidable surgical stent removal  
- stent removal might be difficult in peripheral location | - planned reinterventions to adapt for growth | - high morbidity (operation with CP bypass)  
- higher risk of restenosis compared to endovascular treatment |
Peripheral Pulmonary Artery Stenosis
Adult CHD

• Peripheral PS as part of CHD [TOF / DORV / PA-VSD] or secondary to surgical / interventional therapy

• MRI / CT / catheterization in addition to echocardiogr.

• Indications:
  - RV pressure > 50 mmHg
  - > 50 % diameter narrowing

• Percutaneous interventional therapy is recommended as treatment of choice

• Operations should be performed by surgeons with training and expertise in CHD; multiple thoracotomies markedly increase the surgical risk

Hybrid approach

Hybrid Interventions

• Access to parts of the PA, which cannot be reached by endovascular means
• Enable the use of larger / stiffer catheter / sheath => increasing efficacy / longevity of angioplasty
• less harmful to valves and vessels, avoiding arrhythmia
• Drawbacks: thoracotomy
Peripheral Pulmonary Artery Stenosis
Adult CHD

**Hybrid intervention** for stent angioplasty of peripheral PS:
Lateral thoracotomy for access to peripheral RPA

- PA-VSD after modif. BT-shunt, unifocalization x 2, PKE x 2
- Systemic RV pressure

**Procedure:**
- Sheath in RPA
- Balloon interrogation and stent angioplasty of LPA stenosis
- Stent angioplasty of RPA stenosis
Main pulmonary artery angiogram demonstrating bilateral proximal branch pulmonary artery stenoses (A); simultaneous implantation of stents hand mounted onto 15 mm balloons into the proximal branch pulmonary arteries (B); repeat angiogram following bifurcating stent placement demonstrating widely patent proximal branch pulmonary arteries (C).
Peripheral Pulmonary Artery Stenosis
Adult CHD

Bifurcation stenting II

A: Angiogram demonstrating severe bilateral proximal branch pulmonary artery stenoses and a fractured in situ LPA stent. B: Simultaneous RPA and LPA dilations (12 mm balloons). C: MPA-LPA stent (Intrastent LD Max). D: RPA dilation [4 mm balloon] through the struts of the new LPA stent. E: Simultaneous RPA (12 mm) and LPA (14 mm) balloon dilations. F: MPA-RPA stent. G: LPA dilation [8 mm balloon] through the struts of the RPA stent. H: Simultaneous RPA (14 mm) and LPA (12 mm) balloon dilations. I: Angiogram after intervention demonstrating unobstructed pulmonary arteries.

Peripheral Pulmonary Artery Stenosis
Hybrid Interventions in Infants

- 3 kg, PA-VSD, hypoplastic PA, extreme RV hypertrophy
- s/p RV-PA conduit (8 mm)

Echo: diminutive blood flow from left pulmonary veins
'low output' => ECMO; open chest

Access to LPA with 2.5 coronary button probe and subsequently sheath mounted on this probe
Peripheral Pulmonary Artery Stenosis
Hybrid Interventions in infants

Sheath in RVOT at the prox. end of the RV-PA-conduit

- Stent angioplasty with premounted stent
- favourable angiographic result
Peripheral Pulmonary Artery Stenosis

Follow-up

- All surgical and interventional procedures are burdened by the risk of restenoses:
  - scar formation
  - kinking of large conduits on small PAs
  - tearing / dissection of pulm. arteries
  - thrombus formation
  - "In-stent-stenosis"
  - inefficient radial force of stents
  - outgrowth of stents

- besides echocardiography and cardio-pulmonary exercise test (CPET), additional imaging +/- invasive pressure monitoring is warranted in many pts.

- Lifelong f/u is advised, involving specialists in Adult CHD

Prognosis

Vielen Dank!