What happened to our difficult cases?

Case 4

Bruno Santos Lopes, MD
Department of Cardiology, University Heart Centre, Zurich, Switzerland
**Case 4: N.B., female, 42 years**

**Cardiac anatomy**

**Congenital heart disease:**

- Pulmonary atresia with intact ventricular septum
- Ostium secundum atrial septal defect (ASD II)
Case 4: N.B., female, 42 years

Operations

**Cardiac palliation**

- 1974 (age: 2 days):
  - Brock operation
  - Waterston shunt

**Cardiac repair**

- 1983 (age: 11 years):
  - Take down Waterston shunt
  - RPA reconstruction
  - RV-PA-conduit implantation
Case 4: N.B., female, 42 years
Clinical course until 2009

Complications

- 1994 (age: 20 years): major hemoptysis
- 2005 (age: 31 years): large cavernous pulmonary lesion
Case 4: N.B., female, 42 years
Clinical course until 2009

Complications

- 10/2012: Aspergilloma (*Aspergillus fumigatus*) within the cavernous lesion
- 11/2012: Surgical treatment with atypical right upper lobe wedge resection
Case 4: N.B., female, 42 years
Clinical course 2009/2010

First pregnancy (age: 35 years)

• Hemodynamically well tolerated

• 03/2010: spontaneous delivery with PDA
  • healty girl (birth weight: 2250 g)
  • relevent postpartum hemorrhage with medical treatment only
Case 4: N.B., female, 42 years
At 6 months after delivery in 2010...

Anamnestic and clinical findings

• Mildly impaired physical capacity
• 179 cm, 76 kg, BMI 23.7 kg/m²
• RR 120 / 80 mmHg
• SpO₂ 80-83% (ambient air)
• 2/6 SEM in LUSB
Case 4: N.B., female, 42 years
At 6 months after delivery in 2010…

Anamnestic and clinical findings

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ECG
Case 4: N.B., female, 42 years
At 6 months after delivery in 2010…

Echocardiography
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At 6 months after delivery in 2010…

Echocardiography

CW-Doppler through RV-PA-conduit
Case 4: N.B., female, 42 years
At 6 months after delivery in 2010...

Echocardiography

CW-Doppler through RV-PA-conduit demonstrating restrictive right ventricular diastolic dysfunction with persistent a waves
Case 4: N.B., female, 42 years
At 6 months after delivery in 2010...

Echocardiography

ASD II

L-R-Shunt

R-L-Shunt
**Case 4: N.B., female, 42 years**

*At 6 months after delivery in 2010...*

SpO2 at rest and at peak exercise

<table>
<thead>
<tr>
<th>Date</th>
<th>Rest SpO2 (%)</th>
<th>Exercise SpO2 (%)</th>
<th>Hb (g/dL)</th>
<th>proBNP (ng/L)</th>
</tr>
</thead>
<tbody>
<tr>
<td>07.10.2005</td>
<td>90</td>
<td>-</td>
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**Case 4:** N.B., female, 42 years

At 6 months after delivery in 2010...

Bicycle exercise test

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<tr>
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<tbody>
<tr>
<td>21.03.2005</td>
<td>129</td>
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Case 4: N.B., female, 42 years
At 6 months after delivery in 2010…

Cardiac MRI

- native RVOT: severely stenotic
- extraanatomic anterior RV-PA-conduit: moderate to severe regurgitation (RF 30%)
- stenotic RPA with asymmetrical pulmonary perfusion (RPA : LPA = 16% : 84%)

<table>
<thead>
<tr>
<th></th>
<th>EDVi (ml/m²)</th>
<th>EF (%)</th>
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<tbody>
<tr>
<td>LV</td>
<td>113</td>
<td>40</td>
</tr>
<tr>
<td>RV</td>
<td>normal</td>
<td>57</td>
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Case 4: N.B., female, 42 years
At 6 months after delivery in 2010...

Hemodynamic and angiography

<table>
<thead>
<tr>
<th>CI: 3.6 ml/min</th>
<th>Pressure (mmHg)</th>
<th>SaO2 (%)</th>
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<tbody>
<tr>
<td>SVC</td>
<td>-</td>
<td>60</td>
</tr>
<tr>
<td>MPA</td>
<td>22/8/13</td>
<td>63</td>
</tr>
<tr>
<td>RVSP / RVEDP</td>
<td>52/7</td>
<td>-</td>
</tr>
<tr>
<td>LVEDP</td>
<td>11</td>
<td>-</td>
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<tr>
<td>Aorta</td>
<td>132/62/91</td>
<td>81</td>
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Case 4: N.B., female, 42 years
At 6 months after delivery in 2010…

Hemodynamic and angiography

RV-PA-Conduit regurgitation
native RVOT
RPA stenosis
Result after RPA stenting
Case 4: N.B., female, 42 years
At 6 months after delivery in 2010...

Hemodynamic and angiography

MAPCA (major aortopulmonary collateral artery)
Case 4: N.B., female, 42 years
At 6 months after delivery in 2010...

Hemodynamic and angiography

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<th>During closure</th>
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<td>SaO2 aorta</td>
<td>83%</td>
<td>90%</td>
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<td>RV / RV-EDP</td>
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Transient ASD II closure with sizing balloon
Case 4: N.B., female, 42 years
At 6 months after delivery in 2010...

Hemodynamic and angiography

Transient ASD II closure with sizing balloon

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No permanent ASD closure was performed
Case 4: N.B., female, 42 years
2 years later in 2012…

Anamnestic and clinical findings

- Unchanged mildly impaired physical capacity, riding twice to four times a week
- worked 70%, single mother
- Intermittent palpitations for several minutes as she already had previously

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Case 4: N.B., female, 42 years
2 years later in 2012…

ECG findings

During bicycle exercise test

Holter ECG
Case 4: N.B., female, 42 years
2 years later in 2012…

In summary:

• Patient with repaired pulmonary atresia with intact ventricular septum, 2 years after RPA stenting with unchanged mildly impaired exercise capacity

• Moderate to severe regurgitation and mild stenosis of the RV-PA-conduit

• Restrictive right ventricular diastolic dysfunction with R-L-Shunt through the unrepaired ASD II with increased degree of cyanosis

• Recurrent tachycardias (SVT vs. VT)
Case 4: N.B., female, 42 years
2 years later in 2012...

In summary:

Treatment options:

• Percutaneous pulmonary valve replacement?
• Coiling of MAPCA to the right lung?
• 1 ½ ventricle repair?
• Electrophysiological study?
• ICD?
• Wait and see?
Case 4: N.B., female, 42 years
2 years later in 2012…

Discussion at several interdisciplinary board meetings in Switzerland and at the Advanced Symposium on Congenital Heart Disease in London

Consensus of opinion about treatment

- Percutaneous pulmonary valve replacement
- Electrophysiological study with ablation if possible
- No ICD
- No ASD closure
Case 4: N.B., female, 42 years
From discussion to implementation

Electrophysiological study

- Identification of a monomorphic right ventricular arrhythmogenic focus in the subtricuspid, septal and anteroseptal region with successful ablation

Percutaneous pulmonary valve replacement (Melody®)
Case 4: N.B., female, 42 years
Currently in 2015/2016

Anamnestic and clinical findings

- Stable physical condition, 6 km twice daily with e-Bike
- Works at 90%
- Intermittent premature beats in stressful situations

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<td>65</td>
<td>16.4</td>
<td>227</td>
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<tr>
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<td>89</td>
<td>-</td>
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In conclusion

Patient with repaired pulmonary atresia with intact ventricular septum, severe diastolic right ventricular dysfunction, unrepaired ASD II with bidirectional shunt and intermittent ventricular tachycardias

Almost 4 years after percutaneous pulmonary valve replacement and ablation of right ventricular monomorphic PVC

- stable physical condition
- stable degree of cyanosis
- no arrhythmias

And what about a future pregnancy?
Thank you for your attention