WHAT HAPPENED TO OUR DIFFICULT CASES?

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Lausanne

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RETROSPECT

- 2 years ago a case of a 31 years old female patient was presented at GUCH-colloquium
  - Uncorrected Atrioventricular septal defect (AVSD)
  - Double outlet right ventricle (DORV Fallot type)
  - Trisomy 21
- And the question was late repair YES or NO?
Diagnosis of Atrioventricular septal defect (AVSD) and Double outlet right ventricle (Fallot type) in infancy

Refused for cardiac repair surgery (estimated mortality risk 20%)

1994 – Right Blalock-Taussig shunt

1997 – Modified central Shunt (ascending aorta – MPA) because of thromboed right BT shunt

Stable condition

Last follow up at a cardiologist 1999 (17 years) at University Hospital in Geneva
HISTORY

- 14 years later in 10/2013 treatment in emergency room in Solothurn
- Symptoms of gastroenteritis with diarrhea and vomiting
- Days later increasing abdominal pain ⇒ CT scan
- Thrombosis of the portal vein including confluens of V. lienalis and V. mesenterica superior
HISTORY

- Start Anticoagulation, initial parenteral feeding
- Complicated by acute renal dysfunction (contrast medium-induced) and ascites (Hypalbuminemia) due to liver dysfunction and malnutrition
- Hemodynamical stable, but cyanotic spells with sats 45-50%

Transfer to Inselspital Bern
TRANSTHORACIC ECHO 10/2013

- Complete AVSD
- DORV (Fallot type)
- Moderate AV valve regurgitation
TRANSTHORACIC ECHO 10/2013

- Good biventricular function
- Severe RV hypertrophy
- Gradient across the central shunt 25mmHg
Infundibular RVOT-obstruction
Peak Gradient 63mmHg
Pulmonary valvular annulus 20mm
CT SCAN 10/2013

- Open central shunt (5mm diameter) from ascending aorta to MPA
- 1 small MAPCA
- Good sized MPA, LPA and RPA
- No PA branching stenosis
- 1 small diverticulum without flow
CT SCAN/HOSPITALISATION 10/2013

- Very dilated IVC and liver veins
- Ascites
- Fibro scan negativ
- Discharge home with
  - Marcoumar ⇒ Thrombosis of the portal vein
  - Metoprolol ⇒ Cyanotic spells (severe RVOT-obstruction)
FOLLOW-UP 02/2014

History
- Stable, not very active but not limited (climbing stairs 1-2 flights)
- No syncope or orthopnoe
- Mild ankle oedema with Lasix 20mg od
- No bleeding complications

Clinical findings
- 143cm, 56kg, 27.3kg/m2, Sat 82% at room air
- HF 56bpm, regular, BP right 106/61mmHg, left 122/72mmHg
- RV heave, normal 1.+2. heart beat, 2/6 harsh systolic murmur 2.
  intercostal space LUSB, mild ankle oedema
FOLLOW-UP 02/2014

ECG

- SR, PQ time 280ms (stable with Metoprolol 37.5mg)

Lab

- NTproBNP 644 pg/mL, INR 2.39

Further procedure

- Planned follow-up at hepatology 6 months after thrombosis
- Reevaluation further cardiac diagnostic (heart cath) and rediscussion of option for possible cardiac repair

08/05/2014 – Hypovolemic shock due to upper gastrointestinal bleeding

- Hemoglobin decrease from 170 g/L to 92 g/L
- Transfusion of 3 red blood cell concentrates, 2 FFP
HOSPITALISATION 05/2014

- Gastroscopy
  - 4 esophageal varices (5mm), red signs, fresh blood, Barret esophagus
  - 6x ligations of esophageal varices

- Medical therapy
  - Marcoumar Stop
  - Pantozol, Sandostatine, Ceftriaxon
  - Carvedilol
HOSPITALISATION 05/2014

Complicated course

- Renal dysfunction after CT-scan and anemia ⇒ recovering
- Pulmonary congestion after renal dysfunction and acute volume substitution ⇒ recovering
- Recurrent short episodes with sats 50%
- Under Carvedilol bradycardia 35/min
  ⇒ Carvedilol Stop due to brady tachy syndrome with arrests
CATH 05/2014

Pressures

- IVC 16mmHg
- Liver vein 16mmHg
- RA/LA 12-14mmHg
- LV 108/12mmHg
- Ao 105/54/74 mmHg (Sat 75%)
- PA 22/17/19mmHg (Sat 64%)
PROBLEMS 05/2014

- Chronic portal vein thrombosis, risk for progression due to increased RA pressure and upper gastrointestinal bleeding due to esophageal varices
  - No surgical option for transjugular intrahepatic portosystemic stent-shunt (TIPS)
  - Risk for hepatic failure was judged to be not high if CVP could be lowered
- Brady tachy syndrome with arrests (max 4.7sec)
  - Needs pacemaker
  - But need for anticoagulation for an endovascular pacemaker system
- Multiple cyanotic spells
  - Betablocker contra-indicated
WHAT WOULD YOU DO?

Possibilities

1. Pericardial pacemaker?
2. Endovascular pacemaker (need for anticoagulation)?
3. Cardiac repair & pericardial pacemaker system?
WHAT HAPPENED NEXT

Together with the patient and their family we decided to do

**Cardiac repair & pericardial pacemaker system**

- Operation on 21.10.2014
  - Atrioventricular septal defect repair with two-patch technique
  - RVOT enlargement
  - PA-patch plasty
  - AP-shunt take down
  - Epicardial CRT-Pacemaker
WHAT HAPPENED NEXT

- Good clinical course
- Rivaroxaban (Xarelto 10mg 1x/d) as a prophylaxis for progression of portal vein thrombosis
- Cardiac rehab in Heiligenschwendi
WHAT HAPPENED NEXT

- One relaps with a esophageal varices bleeding in 12/2014 with deterioration of renal function and ascites
- Hepatological reevaluation
  - No possibility of porto-systemic shunt
  - No possibility of recanalization of portal vein system
  - No possibility of liver transplant (no connecting vessels)
- Xarelto Stop: no chances for recanalization, risk of bleeding
- Periodic gastroscopy and ligation of esophageal varices every 8-10 weeks
WHAT HAPPENED NEXT

Combined intervention in 02/2015

- Percutan ASD-Closure (Amplatzer 10mm Septal Occluder) of residual ASD and transjugular liver biopsy
  - RA pressure 6 mmHg
  - Normal liver pressure gradient (liver vein free 16 mmHg, blocked 21 mmHg, hepativ venous pressure gradient 5)
  - Liver biopsy: no liver cirrhosis
  - No intrahepatic shunts

![Hämodynamik: Wedge-Druck in der linken Lebervene: 22 mmHg.](image)
LAST FOLLOW-UP 01/2016

- Good clinical course, no limitations in everyday life
- Stable ascites and liver situation under regular gastroscopies
- Stable renal situation
- Pancytopenia
  - Hb 94 G/L, Lc 2.2 G/L, Tc 53 G/L 18.03.2016
  - Bone marrow puncture rejected
  - JAK2-Mutation negativ
LAST FOLLOW-UP 01/2016

Last Echo 28.01.2016

- Mild stenosis and regurgitation of both AV-valves
- Small residual VSD
Last Echo 28.01.2015

- Normal biventricular function
- Mild pulmonary stenosis