O ρόλος της μαγνητικής τομογραφίας καρδιάς στις συγγενείς καρδιοπαθείς

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Νοσοκομείο ΑΧΕΠΑ
DISCLOSURES

• No relevant financial conflicts
ACHD POPULATION IS INCREASING...

Prevalence of Adult CHD

- Mild
- Moderate
- Severe

Per Million

- 1940-59
- 1960-79
- 1980-89

Adapted from van der Born et al. AHJ 2012

SCMR
ONGOING CARE OF ACHD

- Survival does not equal cure
- Ongoing morbidity and persistent risk of mortality
- Symptoms occur late – often after irreversible damage
- Need for lifelong surveillance - imaging
WHY CMR IN ACHD?

NO RADIATION

LIMITATIONS

- Irregular Heart Rates (AF)
- Small lesions with erratic motion (e.g. vegetations)
- Implantable devices (pacemakers, defibrillators)
- Underestimation of stenotic gradients

Karamitsos TD, Neubauer S. Prog Cardiovasc Dis 2011
HOW TO IMAGE? PREPARATION

- Detailed history is key
  - Original diagnosis
  - Surgical procedures
  - Metallic implants

- Indication for CMR Examination
  - Diagnosis is usually well established
  - Very specific questions – often quantification
ADULT CHD TO DISCUSS

- Tetralogy of Fallot
- Transposition of the great arteries
  - Mustard or Senning
  - Arterial Switch
- Atrial Septal Defects
- Aorta - Valves
- Fontan circulation
TETRALOGY OF FALLOT

TOF Surgery

Table 1 Structural abnormalities encountered after tetralogy of Fallot repair.

- Secondary to surgical repair
  - Pulmonary regurgitation
  - RVOT scar
  - Ventricular septal defect patch
- Residual or recurrent lesion
  - RVOT obstruction
  - Branch pulmonary artery stenosis
  - Ventricular septal defect
  - Atrial septal defect
- Acquired lesion
  - Tricuspid regurgitation
  - RVOT aneurysm
  - Aortic root dilatation
  - Aortic regurgitation
  - Left ventricle dysfunction
- Associated anomalies
  - Systemic-to-pulmonary artery collateral
  - Aortic branch pattern

RVOT: right ventricular outflow tract.
TOF IMAGING GOALS

- Quantify RV & LV volumes and function
- Quantify pulmonary regurgitation
- Clarify anatomy of RVOT
- Look for myocardial fibrosis
- Aortic dilatation and aortic valve regurgitation

CMR IS THE GOLD STANDARD
MODERATE-SEVERE PR

- Mod-severe PR
  - reg fract\textsuperscript{n} 38%

- Mod dilated RV

- Straight outflow tract with good ‘landing’ zone

- RVOT diam
  - 18-22mm
SEVERE PR, DILATED RVOT

- Repaired Fallot’s
- Severe PR
  - reg fractn ~50%
- Very dilated RV
- Straight outflow tract with good ‘landing’ zone
- RVOT diam too large (30-35mm)
CMR PARAMETERS

– WHAT IS THE MAGIC TRIGGER FOR INTERVENTION?

• No ‘magic’ single value
• RVEDVindex >140-170ml/m², z-score> 4.0
• RVESV index >80-90ml/m²
• RV/LV ED ratio >2.0 ……BUT
• Sensitivity & specificity 60-80%
• Beware pts near the ‘cut-off’ – both CMR & clinical uncertainty
• Surgery carries significant risk
• Other parameters important too
  • RVEF <47%, Symptoms, ? QRS>140-160msec
FALLOT – POST SURGERY COMPLICATIONS
TRANSPOSITION OF THE GREAT ARTERIES (TGA)
ATRIAL SWITCH - POST SURGERY ANATOMY

Mustard/Senning Operation

- Ao: Aorta
- MPA: Main Pulmonary Artery
- LA: Left Atrium
- RA: Right Atrium
- RV: Right Ventricle
- LV: Left Ventricle
- SVC: Superior Vena Cava
- Pulmonary Artery
- Pul Veins
- Pul venous pathway
- IVC: Inferior Vena Cava

Ake Senning

William Thorton Mustard
LONG TERM PROBLEMS
POST ATRIAL SWITCH

- Systemic RV failure
- Tricuspid regurgitation
- Venous baffle obstructions
  - Systemic
  - Pulmonary
- Venous baffle leak

IMAGING GOALS

- Quantify systemic RV function
- Evaluate for tricuspid regurgitation
- Assess venous baffles for stenosis or leak
TGA - POST ATRIAL SWITCH

4-CH VIEW – PULMONARY VENOUS RETURN

SYSTEMIC BAFFLE
TGA – POST ATRIAL SWITCH
ARTERIAL SWITCH

**Initial interventions**
- Switch of aortic and pulmonary root
- Anterior positioning of distal MPA / branch PAs (LeCompte manoeuvre)
- Translocation of coronary arteries from aorta to neo-aortic root

**Late interventions**
- AV replacement
- Ao root replacement
- RVOT enlargement
- PA stenting

**Post-operative complications**
- RVOT obstruction / MPA and branch PA stenosis
- LVOT obstruction
- Neo-aortic root dilatation
- Neo-aortic valve regurgitation
- Neo-pulmonary valve regurgitation
- Coronary artery stenosis
- Systemic RV dysfunction
TGA – POST ARTERIAL SWITCH
Coronary artery abnormalities

Pulmonary artery compression or stenosis

Neo-aortic root dilatation

Aortic regurgitation

Mildly narrowed LPA
Atriopulmonary connection  Intracardiac total cavopulmonary connection (lateral tunnel)  Extracardiac total cavopulmonary connection

EXAMPLE – LATERAL TUNNEL FONTAN

- Double-inlet LV with large muscular VSD, two roots, normal great vessels
- Pulmonary artery banding (7 months old)
- Fontan lateral tunnel with 5mm fenestration (18yrs old)
- Recent TIA
- Not compliant with warfarin
EXAMPLE – LATERAL TUNNEL FONTAN
ASD – ANATOMY & FLOW
ASD – PERCUTANEOUS CLOSURE?
EBSTEIN DISEASE

- Άρρεν ασθενής 18 ετών
- Νόσος Ebstein της τριγλώχινας βαλβίδας & Μεσοκολπική επικοινωνία
- Επέμβαση Glenn (6 ετών)
- Σύγκλειση ASD με συσκευή Amplatzer (12 ετών)
- Κολπικές αρρυθμίες & Εύκολη κόπωση
- Echo: σοβαρή Ebstein, μικρή RV, μέτρια TR
- CMR scan to assess further
EBSTEIN DISEASE – GOOD RV SIZE
# Volumetric Data

<table>
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<tr>
<th></th>
<th>ΔΩ (mL)</th>
<th>ΣΩ (mL)</th>
<th>ΟΠ (mL)</th>
<th>ΚΕ (%)</th>
<th>Μάζα (g)</th>
<th>Μάζα/BSA (g/m²)</th>
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<tbody>
<tr>
<td>ΑΚ</td>
<td>137 (102-208)</td>
<td>68 (18-82)</td>
<td>69 (74-150)</td>
<td>50 (57-81)</td>
<td>72 (81-165)</td>
<td>45 (45-81)</td>
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<tr>
<td>ΔΚ</td>
<td>194 (124-256)</td>
<td>92 (38-118)</td>
<td>103 (75-151)</td>
<td>53 (47-71)</td>
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SUMMARY

- Expertise in CMR and familiarity with ACHD is necessary
- A baseline study is useful
- Serial follow up may be needed (no radiation)
- CMR provides unique information in patients with ACHD
  - Clarification of anatomy, functional data, quantification
  - Diagnostic and prognostic data
  - Tissue characterisation