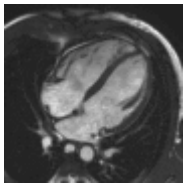


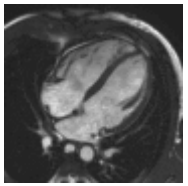
Semmelweis Egyetem
KARDIOLÓGIAI KÖZPONT

XVIII. Ifjúsági Kardiológus Napok

Felnőtt Ivemark szindrómás nőbeteg szív MR vizsgálata

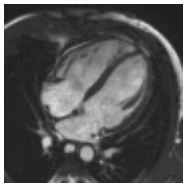
2011, Lillafüred

G.B. 30 éves nőbeteg



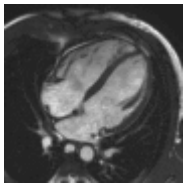
- Ivemark syndroma
- Bal oldali Blalock-Taussig shunt
- Kawashima műtét cyanosis miatt
- Tehermentesítő shunt a bal fülcse és a VCS között
- ECHO: emelkedett pulmonalis nyomás
  - Pulmonalis erek nem voltak láthatók
- Kérdés: anatómiai viszonyok?

Ivemark



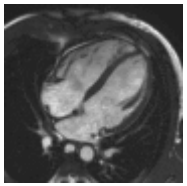
- Jobb pitvari izomerizmus
  - Heterotaxiák közé tartozik (situs ambiguus)
  - Érintett szervek: szív, tüdő, vesék, lép...
    - Asplenia, középvonali máj, GI malrotáció
    - Jobb morfológiájú tüdő és légutak mindkét oldalon
  - Bal isomerismus: polysplenia, bal tüdő
- Svéd patológus írta le a jobb izomerizmust
  - Björn Ivemark (1925 – 2005)

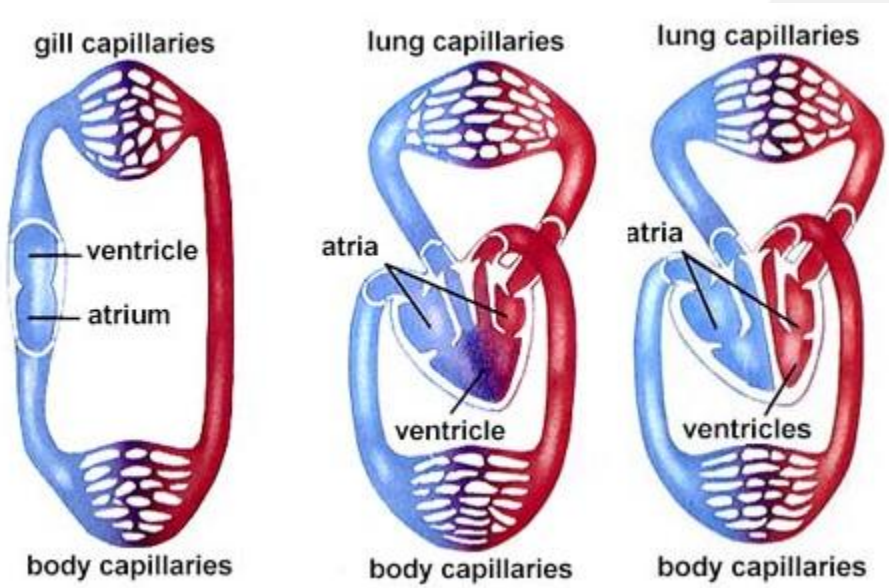
Beavatkozások

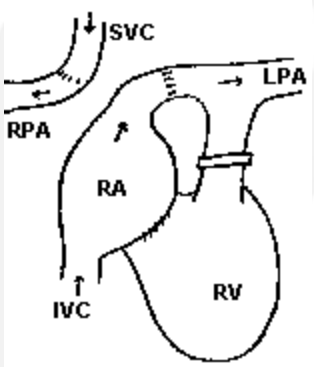


- Blalock-Taussig shunt
  - Arteria subclavia és pulmonalis artéria között
    - Klasszikus: maga a subclavia, végleges palliáció
    - Módosított: műeret használ, ideiglenes megoldás
- Kawashima eljárás – univentricularis szív
  - Bidirectionalis Glenn shunt helyett
    - TCPC (total cavo-pulmonary connection) előtt
  - Vena cava interruptio – azygos continuitas
    - Vena hepaticak változatlanul a pitvarba nyílnak

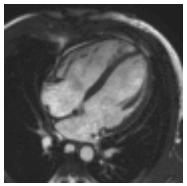
Egykamrás szív: Fontan

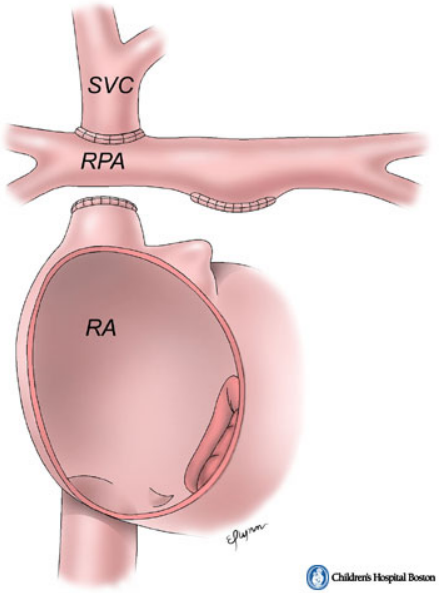


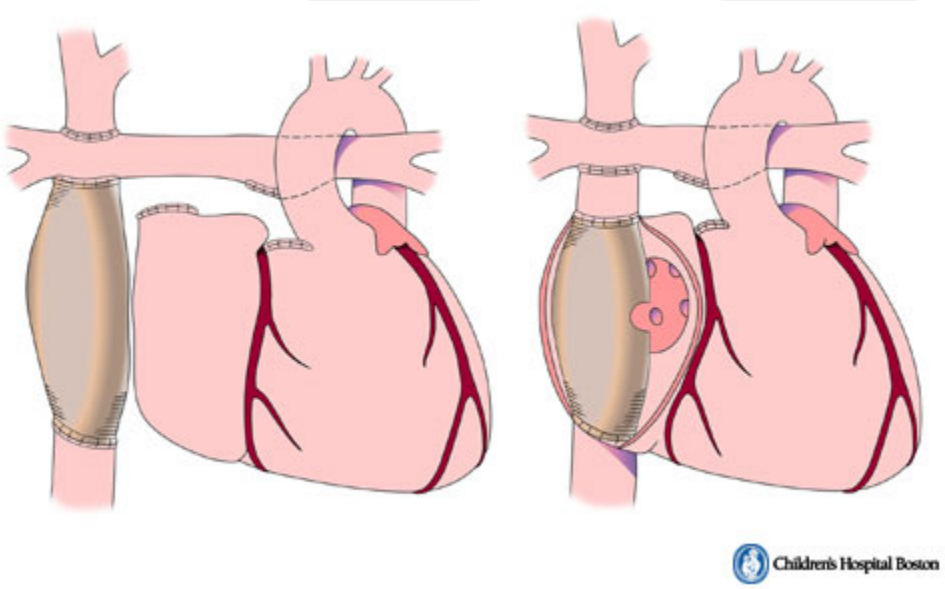




Bidirectionalis Glenn és TCPC







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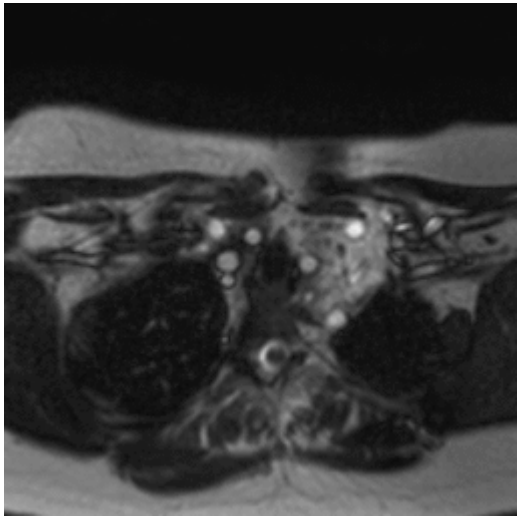
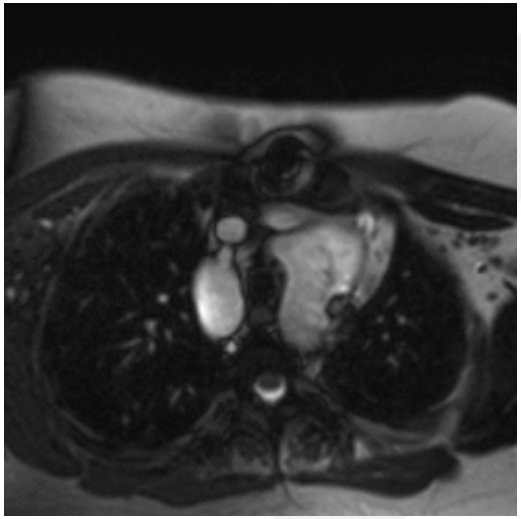
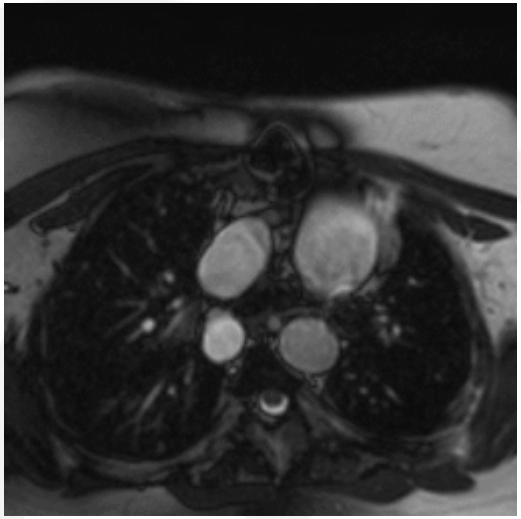
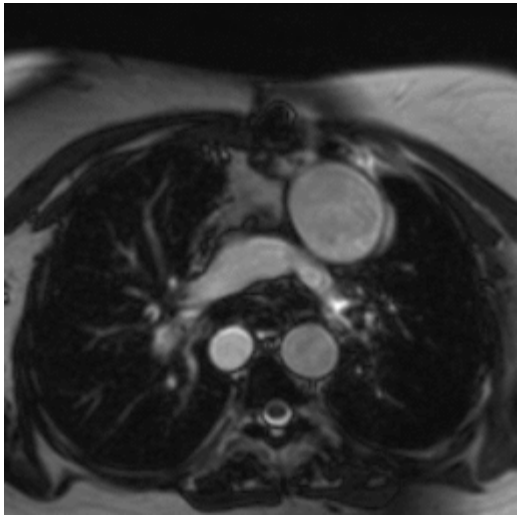
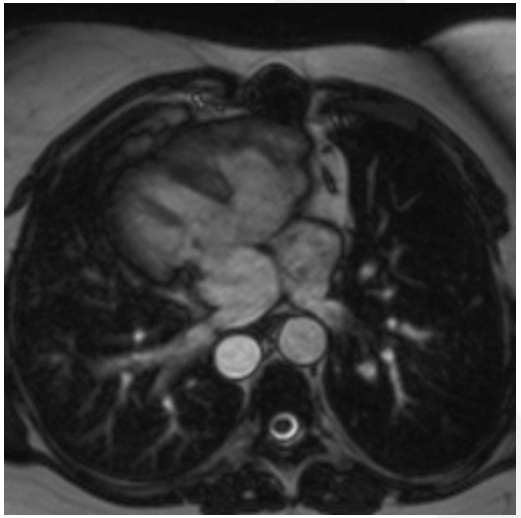
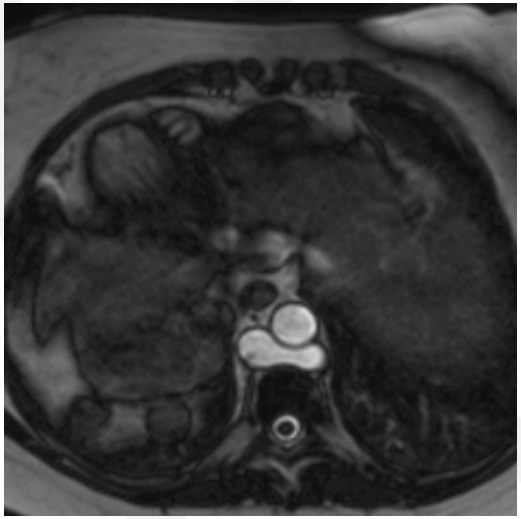

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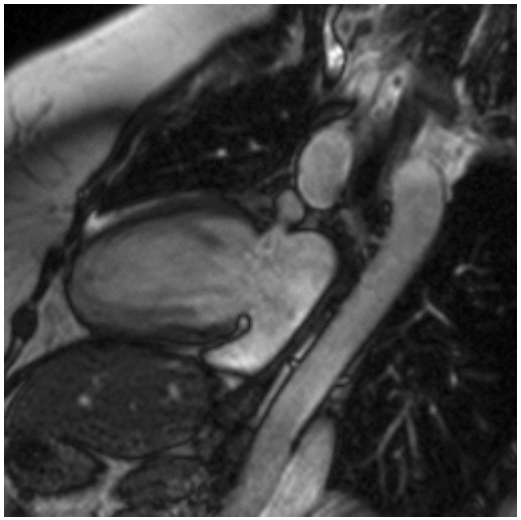
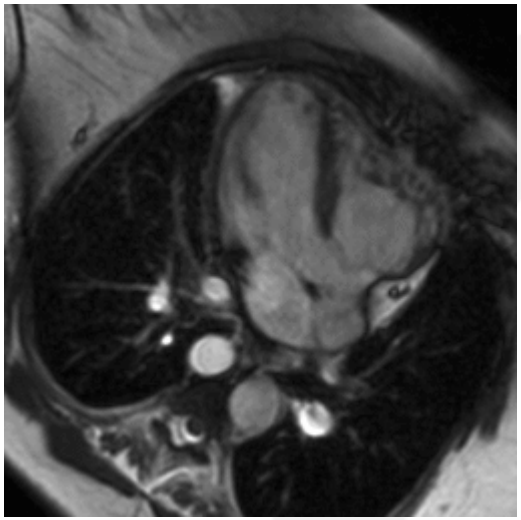
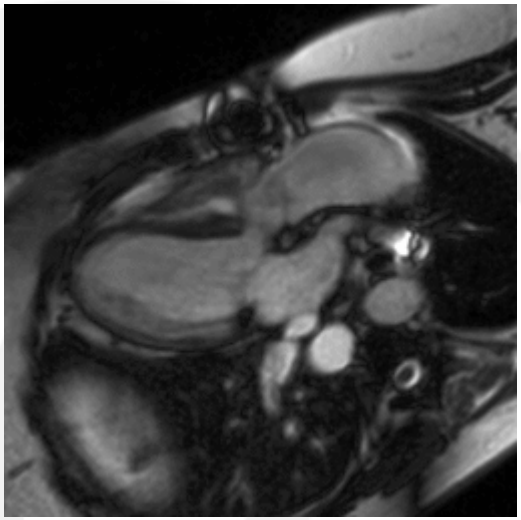
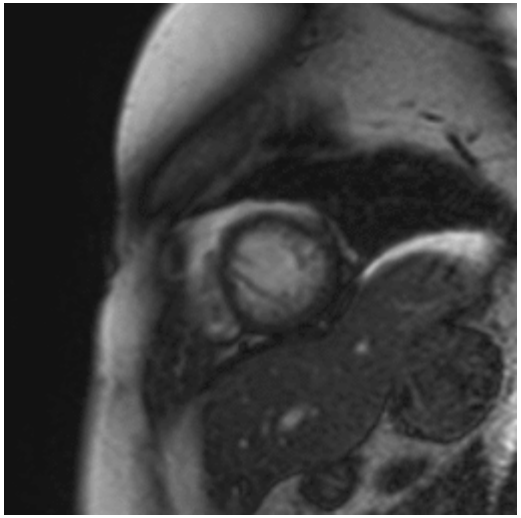
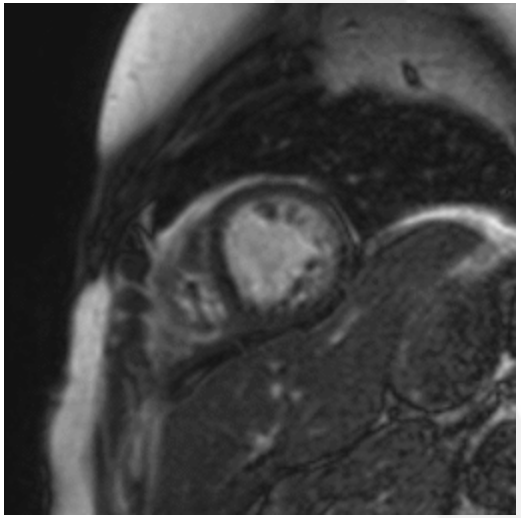
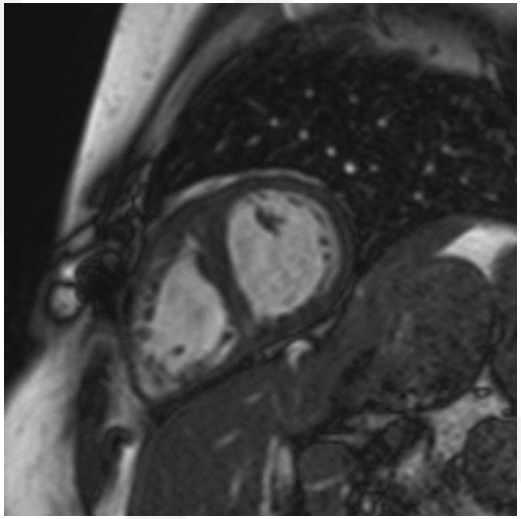
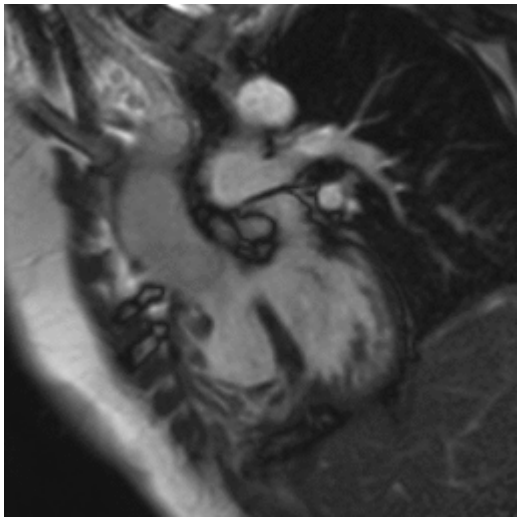
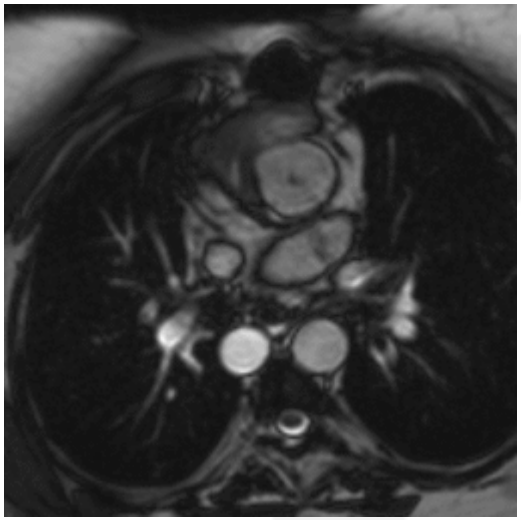
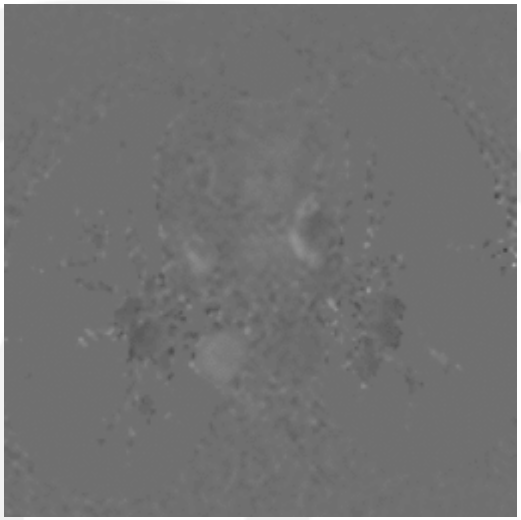
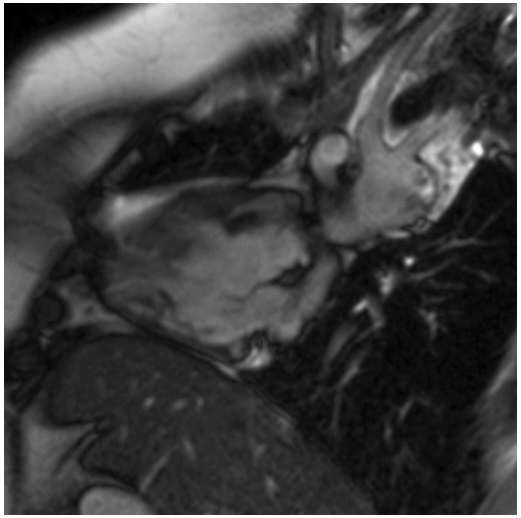
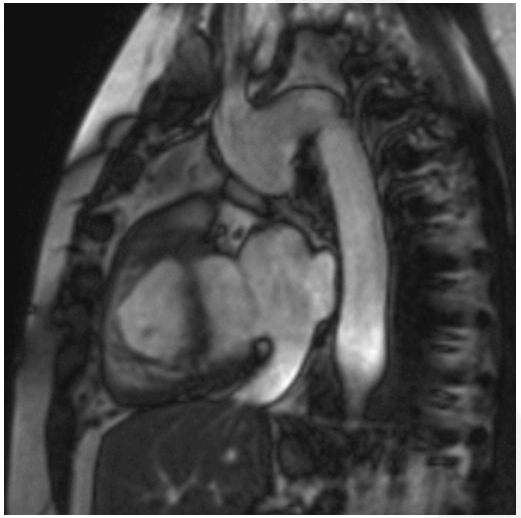
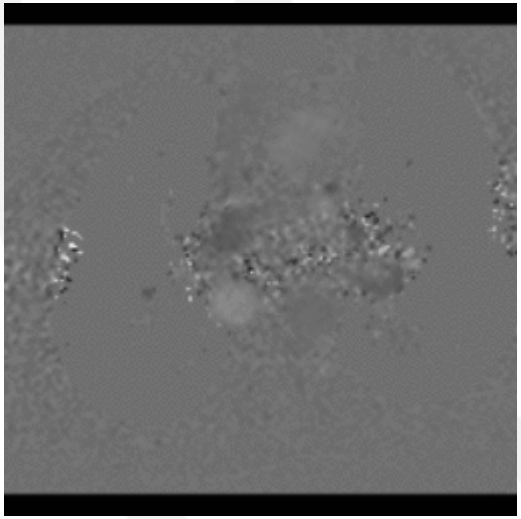
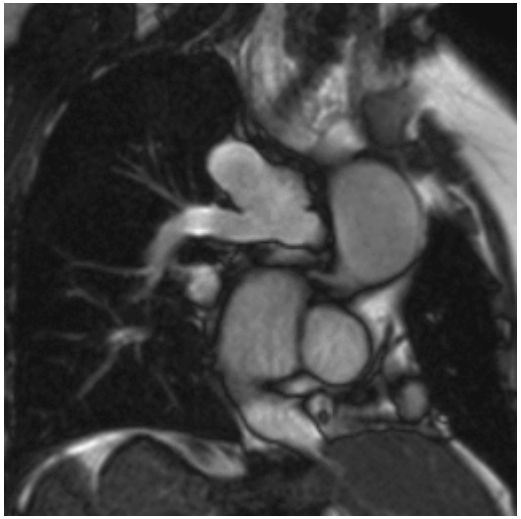
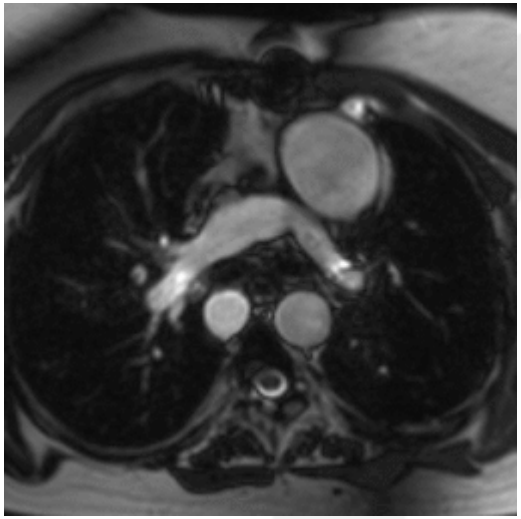
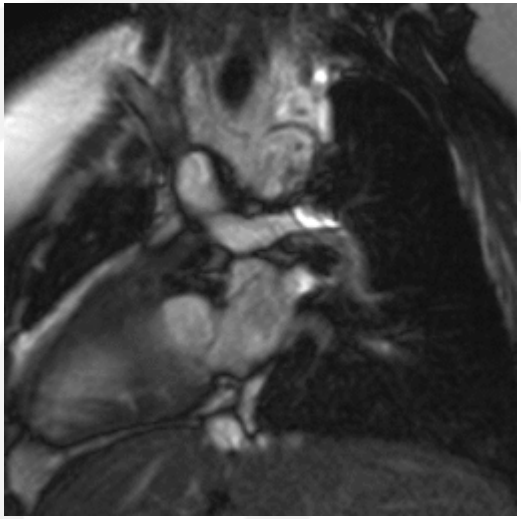
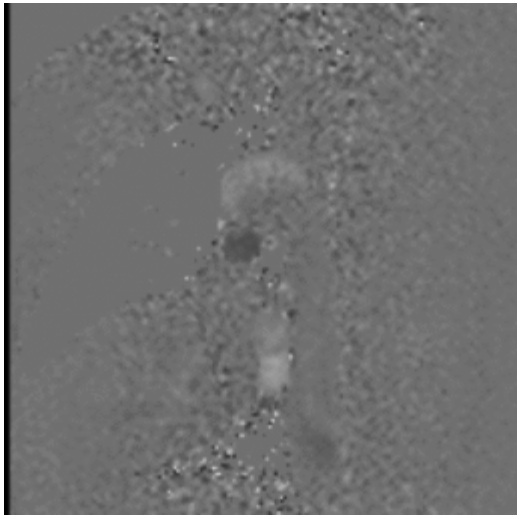
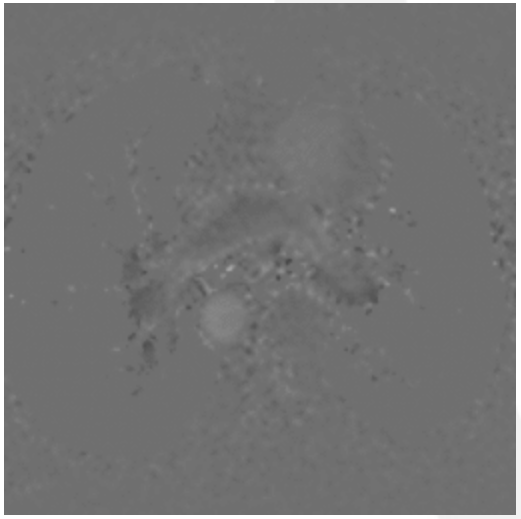
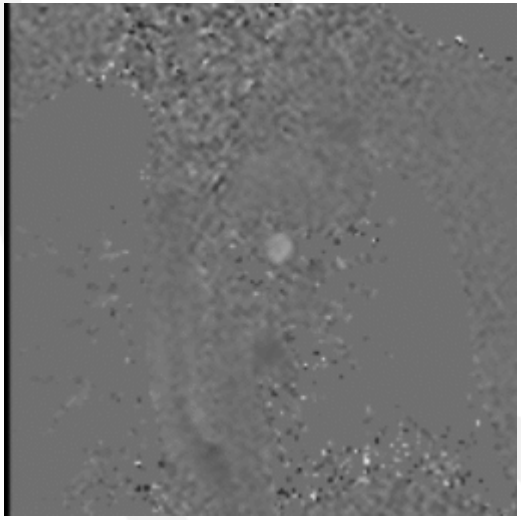







LV: EF 65, ESVi 30, EDVi 85, SVi 55, Mi 59
RV: EF 46, ESVi 34, EDVi 62, SVi 29, Mi 29

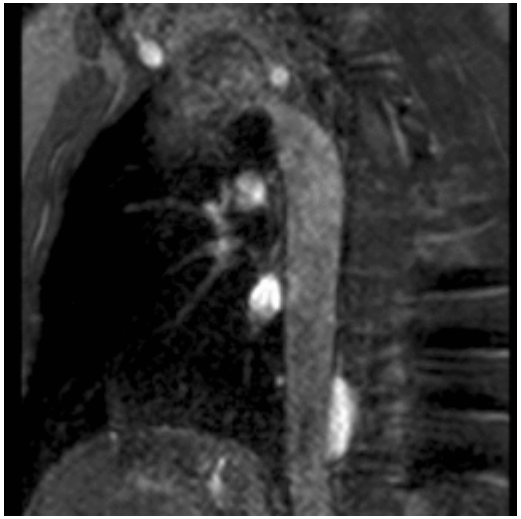
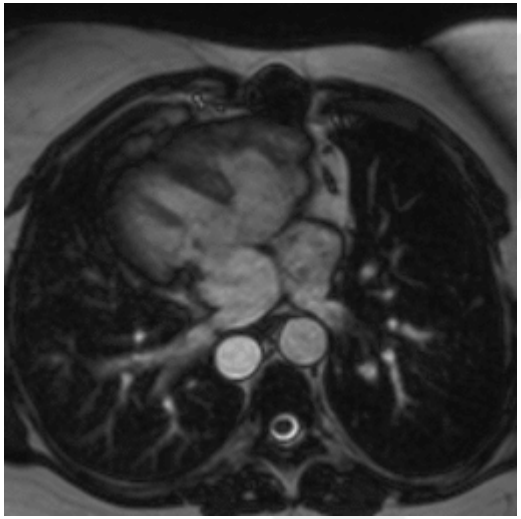
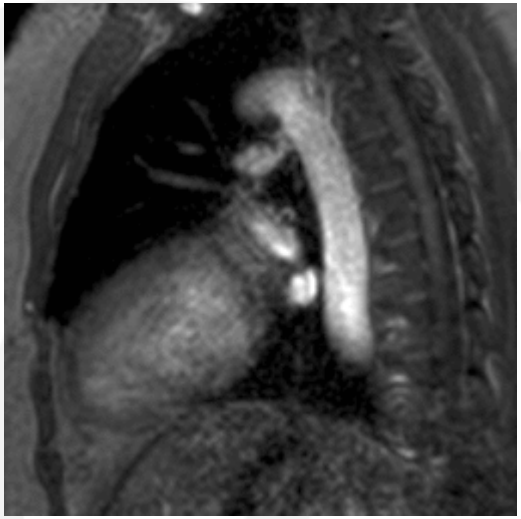
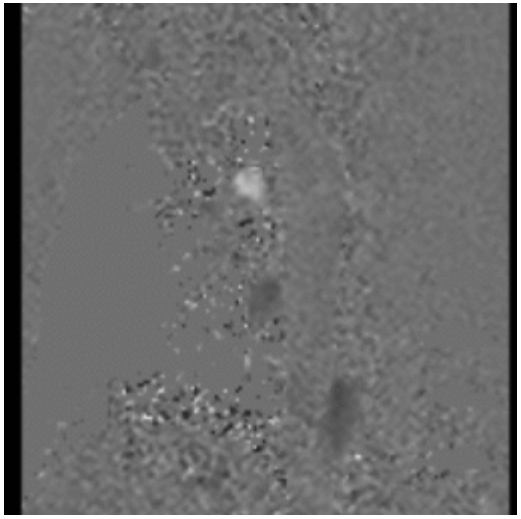
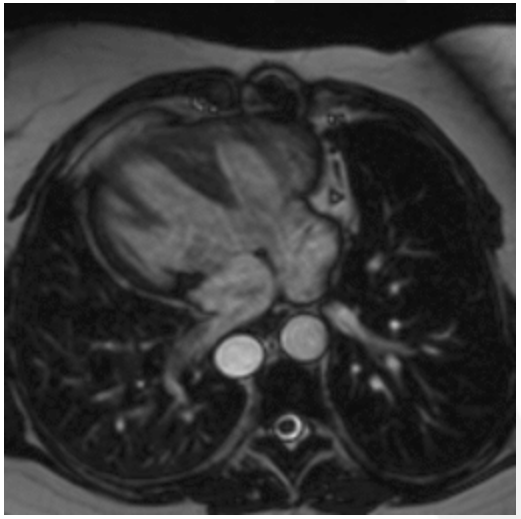
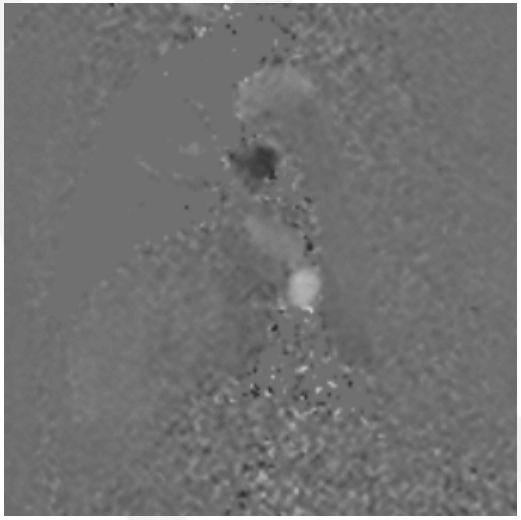
AO: 196cm/s 15Hgmm Rf1%, PA: 1ml

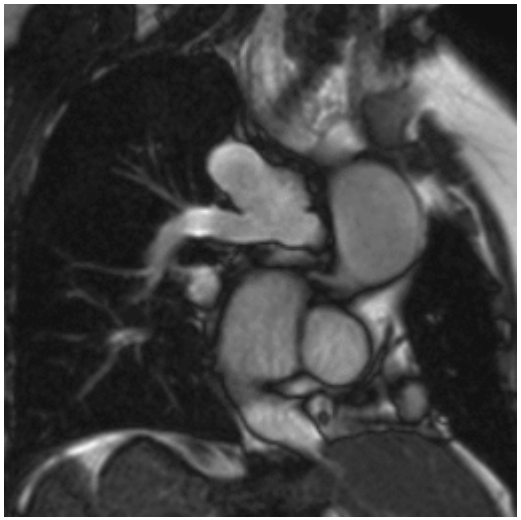
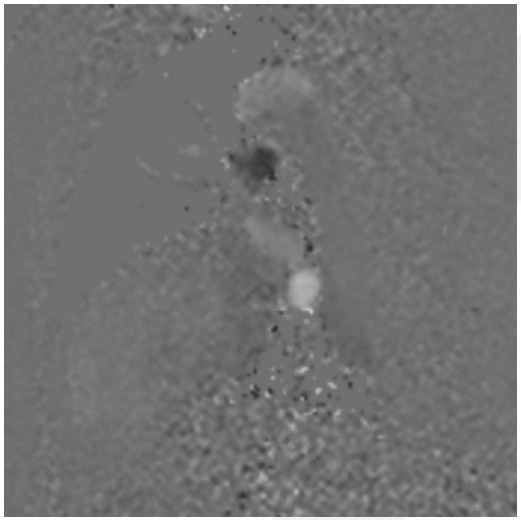
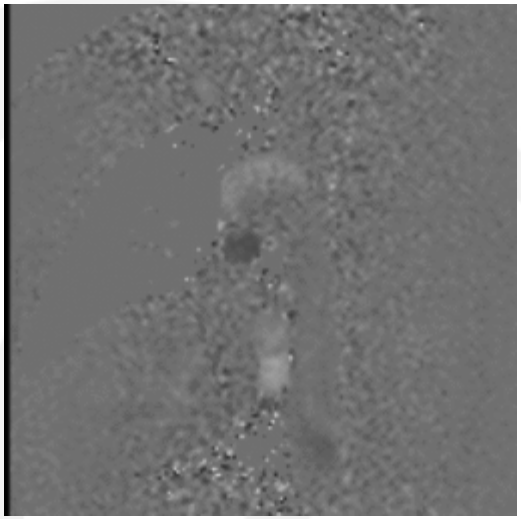
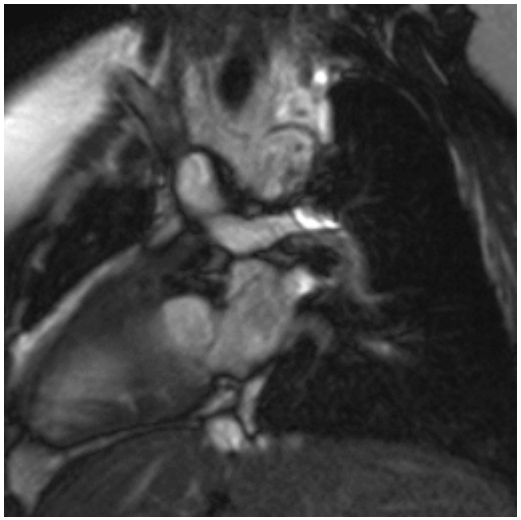
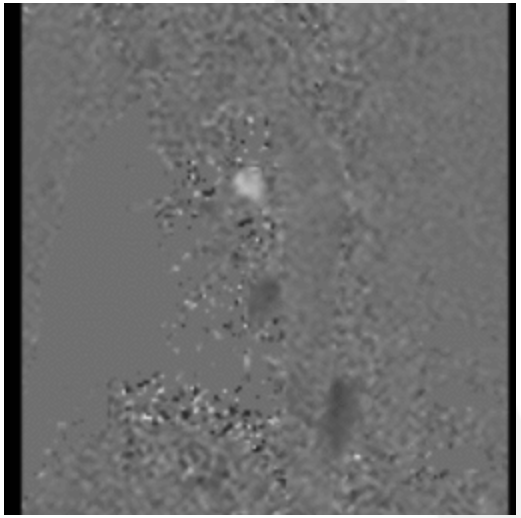
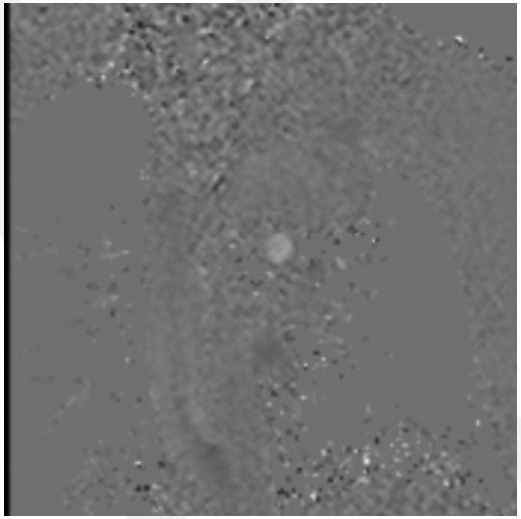
J: 79 3 -, B: 97 4 -, J-B 59-41%

J: 110 5 -, B: 96 3 -, J-B 62-38%

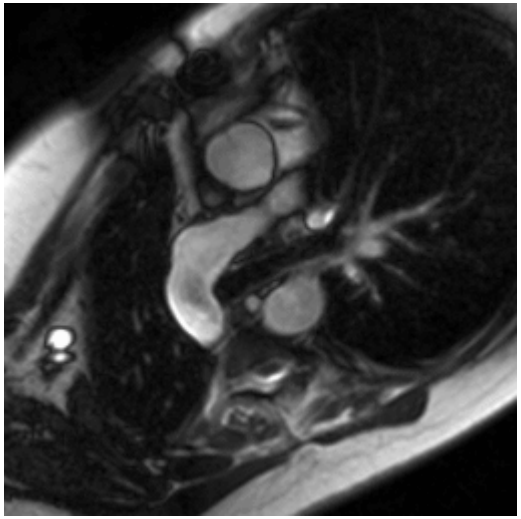
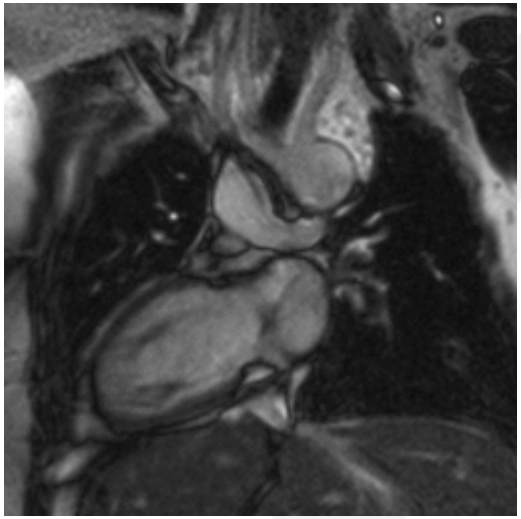
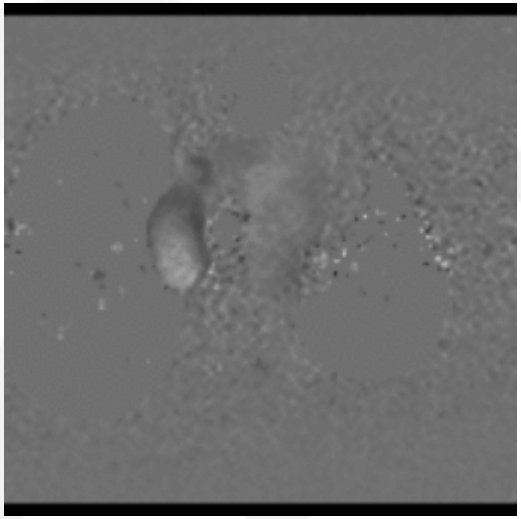
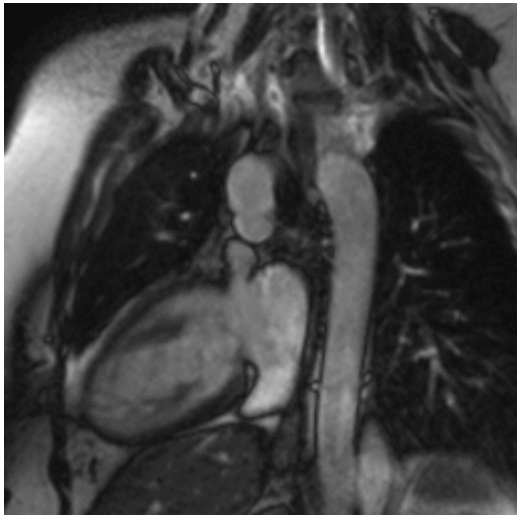
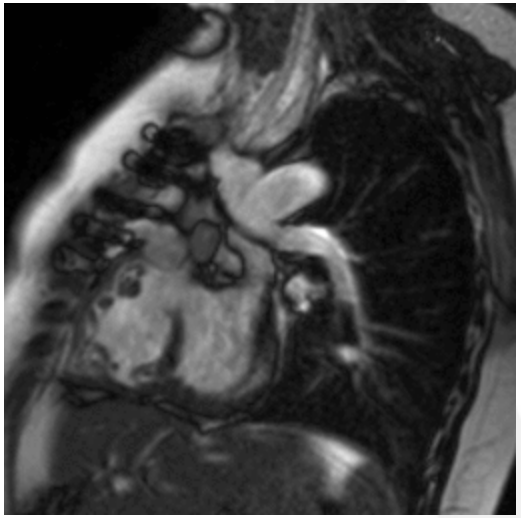
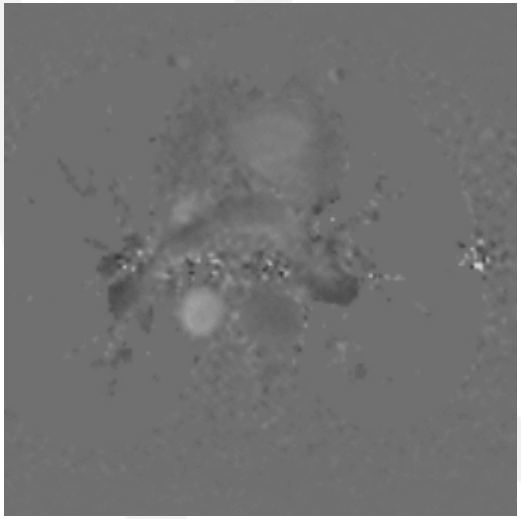







J: 44%, B: 25% micro AVM

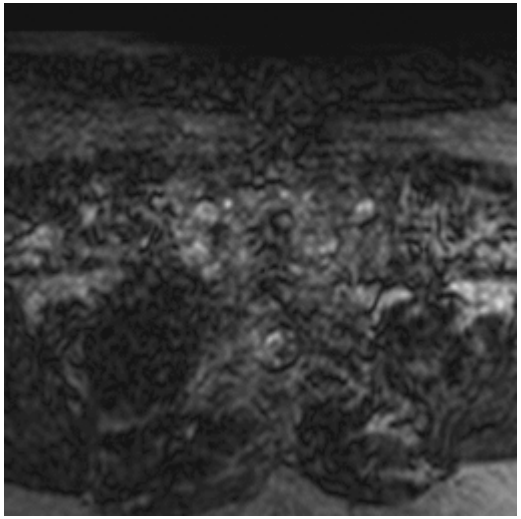
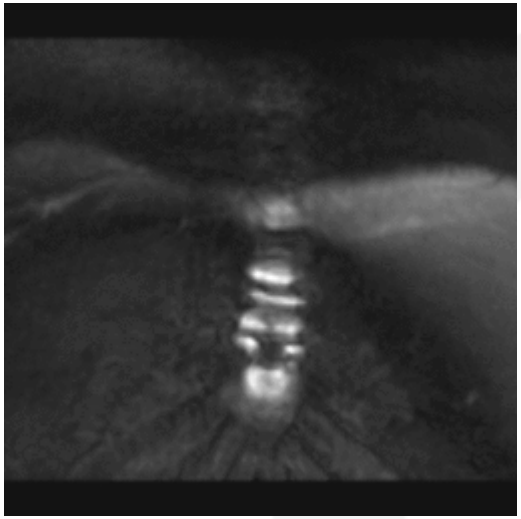
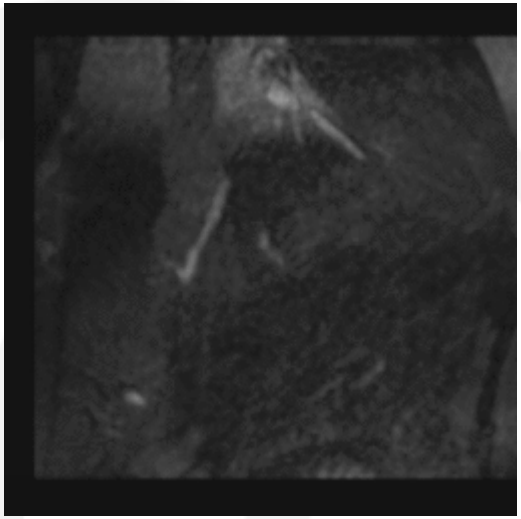
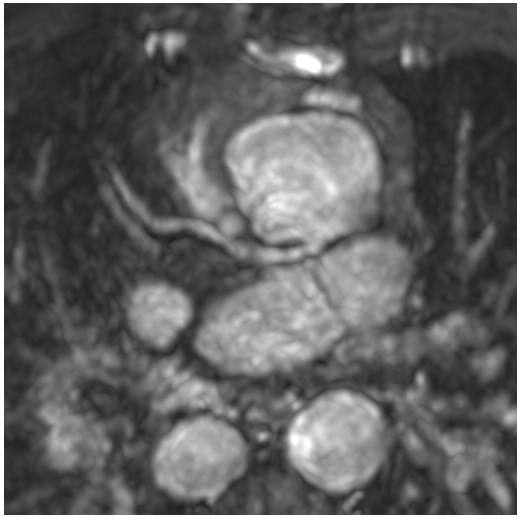
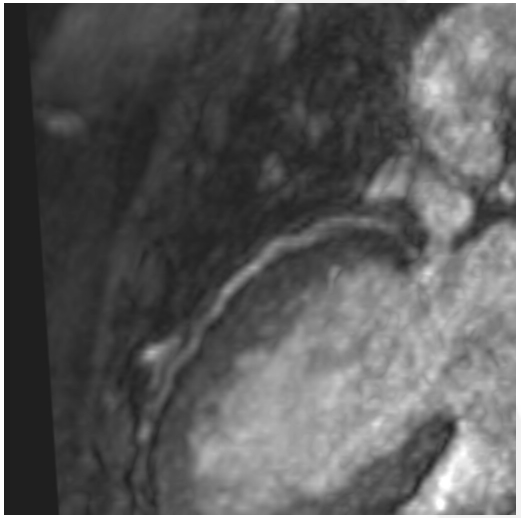
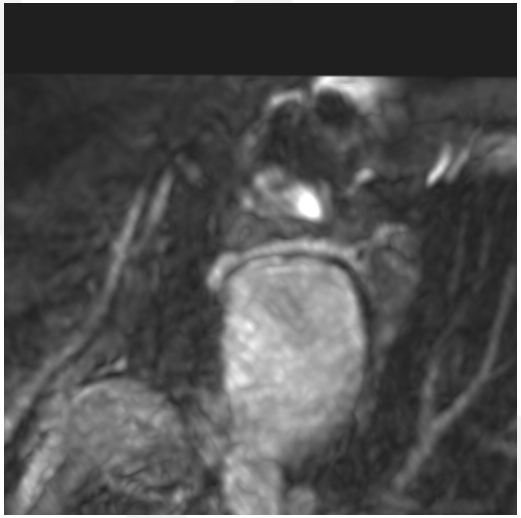







Ann Thorac Surg 80 (5): 1597 – 1603 (2005)

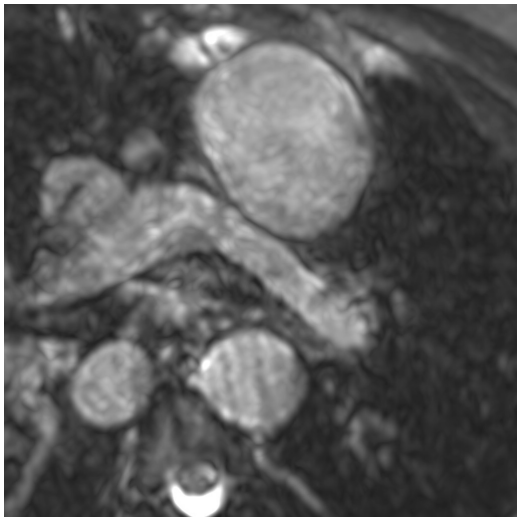
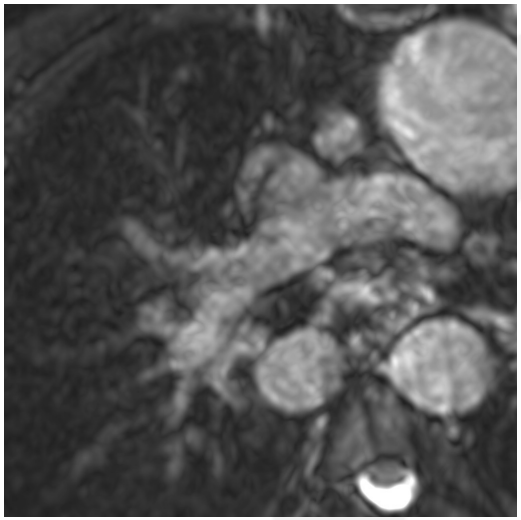
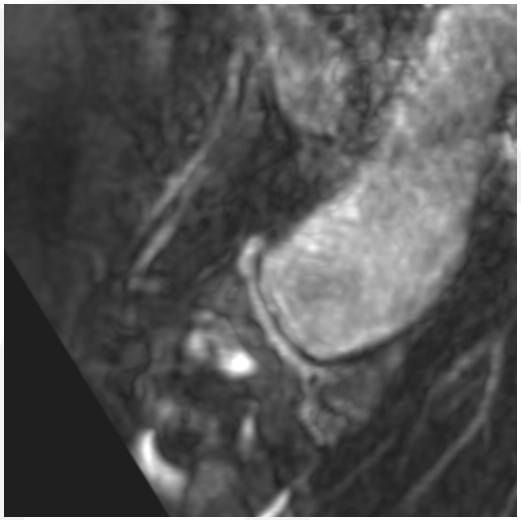
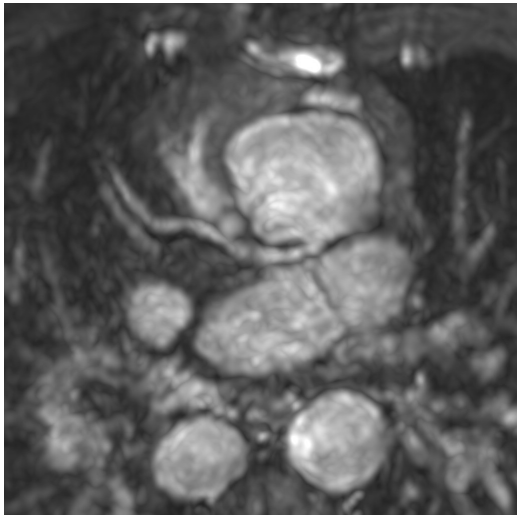
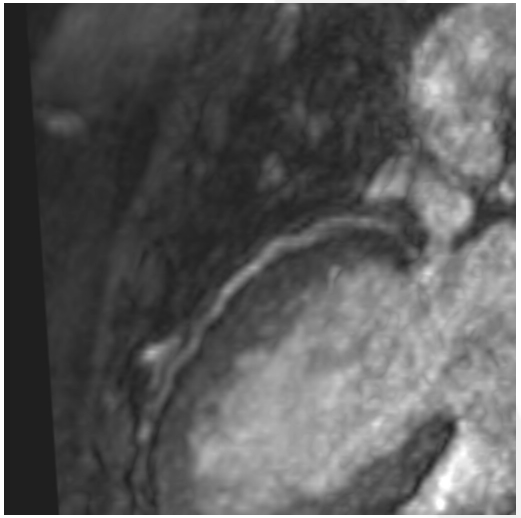
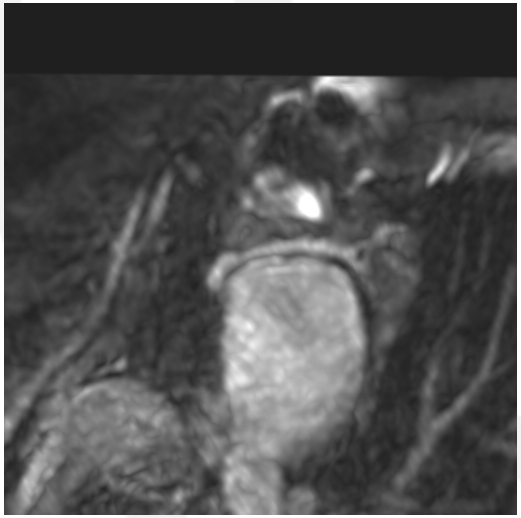
Shunt flow: 2ml, azygos: gyors Fontan

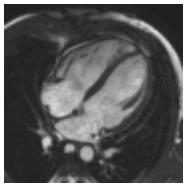
3D NAV MRA

3D NAV MRA

MR eredmény



- Situs ambiguus: bal isomerismus
  - Visceralis situs inversus
  - DORV jelleggel eredő malponált nagyerek
  - A pulmonalis csonkon nincs érdemi leak
  - A fülcse – SVC shunt alig vezet
  - Bal oldalon 25%-os, jobb oldalon 44%-os pulmonalis microAVM shunt
    - Ann.Thorac.Surg. 80(5): 1597-1603 (2005)
  - Ascendenst substernalisan keresztező RCA