Definitions

Unhelpful as guide to venous reconstruction

• Agenesis—IVC (one or all of its segments, called primordia) was never present.
• Aplasia—IVC was present but did not develop normally, perhaps due to in-utero thrombosis.
• Atresia—congenital absence or closure of a normal body opening or tubular organ (Dorland’s); includes agenesis and aplasia.

Disclosures

• Boston Scientific, Medical Advisory Board
• WL Gore & Associates, Consultant

Segmental vulnerability to agenesis and thrombosis are defined by the same large venous confluences.

Thus, iliac vein thrombosis can propagate into the IVC and usually stops at the renal vein confluence.

• 25 year old female with h/o sudden onset of bilateral iliac vein thrombosis in December 2009
• Anticoagulated for one year.
• February 2012, she presented to her primary care with complaints of abdominal pain and leg pain and swelling, and was referred for treatment.
Case lessons

- CT in 2009 shows small-caliber suprarenal and renal IVC present at the time of the acute iliocaval DVT.
- CT shows marked contraction and organization of iliocaval veins, within 3 years. Cord-like veins do not mean agenesis.
- If only the 2012 CT had been obtained, agenesis of the infrarenal IVC would be a natural suggestion.
- Renal and suprarenal IVC agenesis were complicated by infrarenal iliocaval thrombosis. Recanalization must traverse both post-thrombotic and atretic segments.

- 21 y.o. female with history of perinatal cardiac catheterization presented with acute left iliofemoral DVT and failed thrombolysis. CT showed “ageneis” of infrarenal IVC and iliac vein confluence.
Case lessons

• IVC which occludes in perinatal period
  - grows in length proportionately with the rest of the body
  - maintain its structural elements, allowing it to be dilated to adult dimensions
• If this can happen to a child at +3 months, why not at -3 months?

• 34 year old male with Factor V Leiden, history of “IVC thrombosis”, bilateral leg DVT, varicose veins, and healed right leg ulcer
• 6/30/2016 CT: Absent or inconspicuous IVC and small partially calcified iliocaval confluence and common iliac veins.
• 10/31/2016 Iliocaval recanalization, angioplasty, and stenting.

6/30/2016
Case lessons

- The gradual balloon-like inflation of fusiform potential space is seen with perinatal occlusion rather than adult recanalized thrombosed vein.
- Large diameter vein segments, like partially calcified common iliac veins here, imply thrombosis not agenesis. However, string-like residual channels can be seen in thrombosis or agenesis.

Conclusions

- Embryological segments largely coincide with segments between large confluent venous branches (iliac, renal, hepatic veins)
- Longstanding DVT can undergo astonishing degree of organization and contraction
- The cava thrombosed in perinatal period grows like a normal IVC, reaching adult proportions with everything except a lumen.
- Segmental agenesis and perinatal IVC thrombosis are indistinguishable in adulthood.
- Recanalization of long-standing thrombotic occlusions are possible. As always, long term patency depends critically on inflow