IVC Agenesis: Is This a Real Entity

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Disclosures

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

Distinctions which elude HRCT

Unhelpful as guide to venous reconstruction

- Agenesis—IVC (one or all of its segments) 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.
- Chronic thrombotic occlusion: sometimes evident

Primitive segmentation of the IVC corresponds to segments vulnerable to thrombosis, defined by large venous confluences.

Segmental (e.g. infrarenal) IVC thrombosis will parallel embryological segments and can simulate IVC agenesis or aplasia.

In addition, IVC occlusion is a risk factor for leg DVT, so clinical cases may be a mix of atresia and post-thrombotic fibrosis.

- 25 year old female with h/o sudden onset of inferior vena cava clot in December 2009
- Risk factor: OCP.
- Anticoagulated for one year.
- February 2012, she presented to her primary care with complaints of abdominal pain and leg pain and swelling.
The tags "agenesis" and "post-thrombotic fibrosis" are not helpful.

- Agenesis can be complicated by upstream (ie, caudal) thrombosis, as here
- Recanalization may have to traverse both post-thrombotic and atretic segments

If only the 2012 CT had been obtained, agenesis of the infrarenal IVC would be a natural suggestion.

CT shows astonishing degree of contraction and organization of which the iliac veins are capable.
• 21 yo woman, left flank pain, DVU showed acute left iliofemoral DVT from the left common iliac to the popliteal vein, failed thrombolysis. CT showed “agenesis” of infrarenal IVC and iliac vein confluence and thrombus in L ovarian vein.
• History of congenital heart disease involving the pulmonary valve, with perinatal transfemoral cardiac catheterization.

• Suprarenal IVC small caliber but improved
• Low volume collaterals persist
• Patient asymptomatic, but even in 2009, prior to iliocaval DVT, she had evidence of iliportal collaterals and small suprarenal IVC.
• ? Stent suprarenal IVC
Right external iliac vein simulates absent external iliac artery

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

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 and therefore simulate total or segmental IVC agenesis.
• Recanalization of long-standing thrombotic occlusions are possible. As always, long term patency depends critically on inflow

Conclusions
So: IVC agenesis, real or not?

"Doesn't matter." We would attempt recanalization regardless of CT diagnosis and regardless of duration.