18 F-DOPA PET scan for Hyperinsulinism

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What is a PET/CT scan?

Marriage of physiology and anatomy

- Abnormal physiologic activity would be missed by other imaging exams such as MRI, CT, x-ray and ultrasound.
What is a PET scan?

PET = positron emission tomography

- patient is injected with a radiotracer (18F-DOPA)
- the machine picks up the emitted protons in the form of gamma radiation.
A typical PET/CT machine
Dx algorithm

1. Diagnosis of HI
2. 5 day trial of Diazoxide
   - Safety Fast with BS > 70 mg/dL
     - Yes: Diazoxide Responsive
       - Continue Diazoxide
     - No: Diazoxide Unresponsive
       - Stop Diazoxide
         - Initiate glucagon infusion 1mg/day if unable to maintain BS > 70 with dextrose IV
           - Focal: Limited Resection
           - Diffuse: Subtotal Pancreatectomy
3. Send genetic testing
4. Suggests $K_{ATP}$ HI
5. Refer to center with $^{18}$F-Dopa PET Scan

Who benefits from having a PET scan?

- MUST BE PLANNING SURGERY
- Children with one recessive genetic mutation in the KATP channel genes from dad
  - either ABCC8 or KCNJ11 = KATP defect
- Children with no known genetic mutation & who will have surgery (chance that we might find a focal lesion ....low, but not 0)
- Children suspected of having BWS (might be able to have a limited pancreatectomy)
- MUST NOT HAVE GENETICS CONSISTENT WITH DIFFUSE DISEASE!
Why don’t we do PET scans on everyone?

- If child is not going to have surgery, we have no way to confirm if the PET scan was accurate
  - Sensitivity of PET scan is 85%
  - Small focal lesions < 1cm can be missed on PET scan and but identified at surgery.
- Children with HI/HA as these mutations in GDH are known to affect the entire pancreas
- Children with GK HI (again affects the whole pancreas)
- Children with 2 KATP channel mutations, have a dominant mutation, or a maternal mutation are all expected to be diffuse.
Cartoon of focal lesion

Image from Congenital HI video available at http://hyperinsulinism.chop.edu
Focal Pet/CT scan
Diffuse PET/CT scan

Fused image of CT with PET
A variety of focal lesions
The GPS for the surgeon
surgical planning

- If a focal lesion is seen on PET
  - Direct the surgeon to exact location of lesion to limit surgical/anesthesia time.
  - Limit the amount of pancreas removed.
    (preventing future diabetes)
  - Location may be ideal for a laparoscopic procedure.
    (maybe shorter recovery)
ECTOPIC

10 min post injection

Residual head of pancreas

60 min post injection

Ectopic focal lesions

Beckwith-Wiedemann syndrome

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Taking the patient to PET...

24 Hours at CHOP

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Thanks to our multidisciplinary team

Special thanks to study coordinator, Susan Becker RN, who is the “face” of our PET program.