Anatomic Causes for Renovascular Hypertension
Jung-Eun Cheon, MD, PhD

1. What proportion of childhood hypertension is caused by renovascular disease?
   A. 1%
   B. 10%
   C. 30%
   D. 50%
   E. 70%

Correct Answer: B

Rationale(s):
1. Renovascular disease is an uncommon but important cause of childhood hypertension because it is potentially amenable to curative treatment. Renovascular disease causes 5–10% of all childhood hypertension.

Reference(s):

Role of US in Renovascular Hypertension
Ethan A. Smith, MD

2. Which of the following is true of intra-renal resistive indices?
   A. The term parvus et tardus is used to describe normal arterial waveform
   B. The acceleration index is used to evaluate for renal vein abnormalities
   C. Measurement of at least 1 normal value excludes renovascular causes of hypertension

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D. A value of less than 0.5 is abnormal and could indicate a renovascular cause for hypertension
E. Resistive indices should only be measured in the main renal artery

Correct Answer: D

Rationale(s):
1. Normal resistive indices should be above 0.5. Any value under 0.5 in a patient with clinically suspected renovascular hypertension should prompt further evaluation, including possible catheter based angiography.
2. Option A is not correct. Parvus et tardus is used to describe an ABNORMAL arterial waveform.
3. Option B is not correct. Acceleration index is used to evaluate for renal ARTERY abnormalities.
4. Option C is not correct. A single intra-renal RI measurement that is normal does not exclude renovascular hypertension – intra-renal or branch stenosis may result in only small areas of the kidney with abnormal Doppler values. In fact, multiple normal values cannot completely EXCLUDE renovascular hypertension.
5. Option D is not correct. Resistive indices are typically measured within the kidney, not in the main renal artery.

Reference(s):

Role of CT in Renovascular Hypertension
Monica Epelman MD

3. 3-year-old with presents with hypertension and several flat, pigmented spots on her skin and freckling under the arms. A renal CTA image is shown below. What is the most likely diagnosis?
   A. Right renal artery high-grade ostial stenosis in the setting of Neurofibromatosis type 1
   B. Right renal artery high-grade ostial stenosis in the setting of Neurofibromatosis type 2

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C. Right renal artery aneurysm in the setting of Neurofibromatosis type 1
D. Right renal artery aneurysm in the setting of Neurofibromatosis type 2

Correct Answer: A

Rationale(s):
1. Café-au-lait spots, are flat, light tan color, pigmented spots on the skin, which are called by the French term for coffee (café) with milk (lait). These are the most common sign of NF 1 and the presence of 6 or more spots is considered one of the diagnostic criteria for NF1. Café-au-lait spots are usually present at birth or generally appear by two years of age. Freckling under the arms or in the groin area is an additional criteria for NF1. Axillary freckling is not seen in every person with NF, but when present it is considered strong evidence of NF 1. NF1 may be also associated with a mesodermal vascular dysplasia that can affect the abdominal aorta and its branches, most commonly the renal arteries. Potential etiologies of hypertension in children with NF1 include renal artery stenosis and, infrequently, pheochromocytoma. Arterial distribution of NF1 is similar to the vasculopathy seen in patients with atherosclerosis with ostial renal artery stenosis being observed more commonly than intraparenchymal disease. The image above shows high grade right renal artery stenosis with mild post-stenotic dilatation.

2. Option B is not correct. NF2 is characterized by the development of bilateral vestibular schwannomas. It is clinically distinct from NF1 and is ten times less common. Unlike NF1, individuals with NF2 have few skin abnormalities and do not typically exhibit café-au-lait spots, axillary freckling or renovascular disease.

3. Option C is not correct. The image shows high grade right renal artery stenosis with mild post-stenotic dilatation. The image does not show a right renal artery aneurysm.

4. Option D is not correct. The image shows high grade right renal artery stenosis with mild post-stenotic dilatation. The image does not show a right renal artery aneurysm.

Reference(s):

Role of Angiography in Renovascular Hypertension
Derek J. Roebuck, MD

4. Angioplasty is an appropriate treatment for many children with renovascular hypertension. Which statement is correct?
   A. Most children undergoing angioplasty have an underlying syndrome (such as neurofibromatosis type 1).

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B. It is appropriate to maintain blood pressure above the 95th centile for age and height in some children.
C. Angioplasty is technically successful in 98% of cases.
D. About 70% of children with renal artery stenosis are cured by angioplasty.

Correct Answer: B

Rationale(s):
1. A small but significant proportion of children with renovascular hypertension have coexisting cerebrovascular disease. These children may be at risk of neurological symptoms of even stroke if their BP is reduced to normal.
2. Option A is not correct. About 30% of patients treated for renovascular disease have an underlying syndrome.
3. Option C is not correct. Angioplasty is not successful in about 10% of children who undergo angiography – usually due to involvement of a very small artery, or complete renal artery occlusion.
4. Option D is not correct. Probably <30% are cured, but most have improved BP control.

Reference(s):

Role of MRI in Renovascular Hypertension
Franz Wolfgang Hirsch, MD

5. CE-MRA is often performed in children with suspected renal arteries stenosis. Which statement is correct?
   A. The sensitivity and specificity of CE-MRA is as good as of CTA and DSA. This also applies to stenoses of second order branches and third order branches of the renal arteries.
   B. The sensitivity and specificity of CE-MRA is as good as of CTA in detection of renal artery stenosis but this only applies for the main renal arteries.
   C. CE-MRA is well suitable in children with renal insufficiency.
   D. Bolus timing of contrast agent is not crucial in CE-MRA.

Correct Answer: B

Reference(s):

Please return your completed cover sheet and answers to the personnel at the IPR Registration Desk.
Role of US in Renovascular Hypertension
Ethan A. Smith, MD

6. Which of the following is true regarding renal Doppler US?
   A. Asymmetric renal size is the most specific finding indicating renovascular causes of hypertension
   B. Both false negatives and false positives occur, so further imaging is needed in patients with clinically suspected renovascular hypertension even if the ultrasound is normal
   C. Excellent sensitivity for accessory renal arteries
   D. Less sensitive, but more specific than catheter based angiography
   E. Only color or power Doppler images are needed when evaluating for causes of renovascular hypertension

Correct Answer: B

Rationale(s):
1. Both false negatives and false positives occur, so further imaging is needed in patients with clinically suspected renovascular hypertension even if the ultrasound is normal.
2. Recent studies have shown that renal ultrasound with Doppler performs better than historically reported, but that both false negatives and false positives still occur. Therefore, if renovascular hypertension is suspected based on the patient's clinical presentation, further evaluation with CTA/MRA or catheter based angiography are needed.
3. Option A is not correct. Asymmetric renal size could indicate a vascular abnormality, but can also be seen with reflux nephropathy and other non-vascular causes.
4. Option C is not correct. One weakness of ultrasound is the difficulty identifying small accessory renal arteries.
5. Option D is not correct. Ultrasound with Doppler is less sensitive and less specific than catheter based angiography, the gold standard imaging test.
6. Option E is not correct. Spectral Doppler images with evaluation of renal arterial waveforms are critical when evaluating for renovascular causes of hypertension.

Reference(s):

Please return your completed cover sheet and answers to the personnel at the IPR Registration Desk.
Role of CT in Renovascular Hypertension
Monica Epelman MD

7. What is the most likely cause of elevated blood pressure in this 14-year-old female with a past medical history of vesicoureteral reflux?
   A. Fibromuscular dysplasia
   B. Essential hypertension
   C. Renal parenchymal disease
   D. Coarctation of the aorta

Correct Answer: C

Rationale(s):
1. Most cases of hypertension in young children are secondary to an underlying disorder. Renal parenchymal disease is the most common cause of hypertension in children younger than 6 years of age and older than 1. In adolescents, hypertension is usually primary or essential in most instances, making up 85 to 95 % of cases. However, renal parenchymal disease is the third cause of hypertension after iatrogenic illnesses. Other possible etiologies include endocrine disease (e.g., hyperthyroidism, pheochromocytoma) and coarctation of the aorta. In general, the likelihood of identifying a secondary cause of hypertension in children is proportionally related to the degree of blood pressure elevation and inversely related to the age of the child. The image in this case shows right renal scarring and shrinkage characteristic of reflux nephropathy and likely the cause of hypertension in this patient.
2. Option A is not correct. Though fibromuscular dysplasia (FMD) may cause of renovascular hypertension in this patient. No imaging signs of FMD are seen in this image and FMD is not the most cause for hypertension at this age. Furthermore, the renal arteries are not shown.
3. Option B is not correct. Given the imaging findings in the right kidney, hypertension in this patient is most likely secondary to renal parenchymal disease, reflux nephropathy in this case.
4. Option D is not correct. Though coarctation of the aorta may manifest as hypertension in young patients, no imaging or clinical findings suggestive of this diagnosis are provided.

Reference(s):

Please return your completed cover sheet and answers to the personnel at the IPR Registration Desk.
Role of Angiography in Renovascular Hypertension

Derek J. Roebuck, MD

8. With respect to endovascular treatment of hypertension in children:
   A. Most disease involves the aorta and/or main renal arteries.
   B. More than 80% of children will need only one procedure.
   C. It is possible to treat lesions of arteries <1.5 mm in diameter.
   D. Stents should not be used in the renal arteries of children.

Correct Answer: C

Rationale(s):
1. Arteries <1.5 mm in diameter can be treated. Although the long-term results of angioplasty in these small branches are unknown, ethanol ablation may be effective.
2. Option A is not correct. Only a minority of lesions involve the main renal arteries.
3. Option B is not correct. Somewhere between 20% and 40% of children will need more than one procedure.
4. Option D is not correct. Stents are indicated in some circumstances, for example following recanalization of longstanding renal artery occlusion.

Reference(s):

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9. Which of the following is not a condition associated with renal artery stenosis?
   A. Edward’s syndrome
   B. Moyamoya disease
   C. Williams’ syndrome
   D. Tuberous sclerosis
   E. Neurofibromatosis type 1

Correct Answer: A

Rationale(s):
Several rare syndromes or conditions are associated with childhood renovascular hypertension, the most common being neurofibromatosis type 1. The prevalence of renal...
artery stenosis in patients with moyamoya disease was 5%

Reference(s):