

SPR 2014 Interventional Session
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SAM Questionnaire

Management of Benign Biliary Strictures

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1. What anomaly is always associated with benign biliary obstruction?

- A. Cholestasis and or cytotoxicity
- B. Bile duct dilatation on imaging (US, CT MRI)
- C. Jaundice and/or pruritus
- D. None
- E. Abdominal pain

Correct Answer: D

References

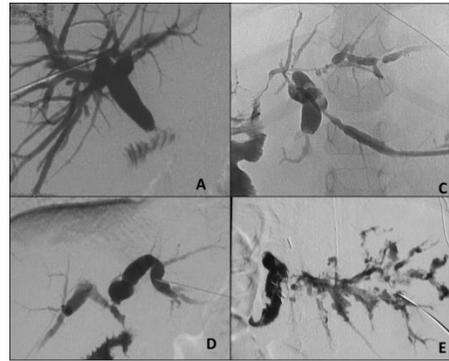
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Rationales

- A. Partial biliary obstruction secondary to benign stenosis or stone, may present with normal hepatic biology especially if the patient is treated with ursodeoxycholic acid
- B. B: The correlation between the degree of bile duct dilatation and the severity of biliary obstruction is poor. Even in the presence of severe stenosis, bile duct may not be enlarged making the access for biliary transhepatic intervention difficult.
- C. C: As answered for A and B, correlation between severity of biliary obstruction and clinical signs can be poor.
- D. D: Significant biliary obstruction can evolve with no clinical, biological or imaging call signs. Because of the chronic bile obstruction, fibrosis can occur and even evolve to cirrhosis. Functional imaging such as elastometry could help in detecting an abnormal evolution in a patient presenting otherwise normal follow-up
- E. E: Abdominal pain is usually associated with acute obstruction due to bile stones. It is rare in infants presenting stones and in patients presenting post-surgery progressive stenosis.

2. After liver transplantation, what type of stenosis has the best prognosis?

- A. Isolated anastomotic stricture (bilio-enteric or bilio-biliary)
- B. Multiple intrahepatic strictures
- C. Multiple intrahepatic strictures associated with cholangitis
- D. Anastomotic and intrahepatic stenosis
- E. Multiple intrahepatic strictures associated hepatic artery thrombosis



Correct Answer: A

References:

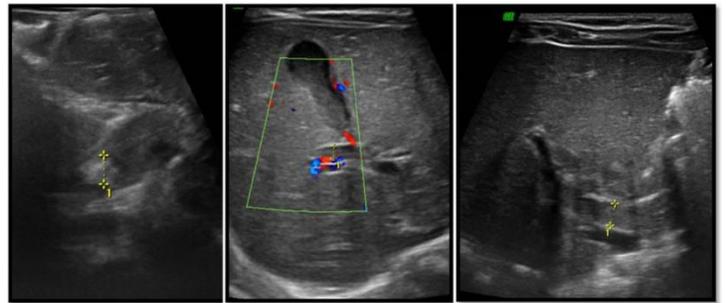
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Rationales

- A. Simple anastomotic strictures post Liver Transplantation has the best success rate after percutaneous ranging from 58 to 100 % according to the reported series. In case of failure of the percutaneous treatment these strictures can be treated surgically
- B. Intrahepatic strictures have a bad prognosis as only interventional biliary procedure can treat them but with a low success rate .
- C. Cholangitis may mimic multiple intrahepatic stenosis that can completely or partially disappear after antibiotherapy +/- external biliary drainage
- D. The prognosis of intrahepatic stenosis associated with anastomotic stricture is related to the presence of intrahepatic strictures
- E. Hepatic artery thrombosis that lead to biliary ischemia and post-ischemic cholangiopathy has a poor prognosis

3. A 12 day old boy presents with acholic stools, jaundice and cholestasis and cytotoxicity on blood test. Here is the aspect on ultrasonography. What are your diagnosis and treatment?

- A. Biliary atresia and surgery
- B. Cystic fibrosis and medical treatment
- C. Choledocal cyst and surgery
- D. Biliary lithiasis and surgery
- E. Biliary lithiasis and percutaneous treatment



Correct Answer: D

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Rationales

- A. Intrahepatic bile duct dilatation is the only sign that permits to rule out biliary atresia. A normal liver ultrasonography is possible at the early stage of biliary atresia and does not permit to rule out the diagnosis.
- B. Cystic fibrosis may cause cholestasis with acholic stools but is not associated with bile ducts dilatation.
- C. Choledocal cyst is usually larger than the common bile duct seen on this slide, but it can be associated with bile stones secondary to bile obstruction and mimic “simple” lithiasis obstruction. This diagnosis can be ruled out by showing a normal bilio-pancreatic junction length (<5mm) during percutaneous cholangiography or on MR-Cholangio-Pancreatography.
- D. Biliary obstruction secondary to biliary lithiasis. . Surgery should be reserved to percutaneous treatment failure
- E. Biliary obstruction secondary to biliary lithiasis. If the obstruction does not resolve spontaneously within a two weeks delay or if signs suggestive of cholangitis occur, percutaneous treatment should be performed.

Slow Flow Vascular Malformations

John J. Crowley

4. The inheritance of Blue Rubber Bleb Nevus Syndrome (BRBNS) is:

- A. Autosomal dominant
- B. Autosomal recessive
- C. Sex linked recessive
- D. Sporadic

Correct Answer: Answer is D: There are no well documented cases of heritable BRBNS. Recent genetic analysis of a cohort of BRBNS patients has demonstrated post zygotic TIE 2 mutations without germline mutations.

References

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5. Features which help distinguish a glomuvenous (GVM) malformation from a “garden variety” venous malformation include:

- A. GVM's are typically nodular and scattered or plaque like and regional, varying in color from pink to purplish dark blue whereas common venous malformations are soft, blue and either localized or diffuse.
- B. GVM's have a distinct raised, often hyperkeratotic cobblestone-like appearance, venous malformations do not.
- C. In contrast to venous malformations, GVM's are painful to palpation, and cannot be completely emptied by compression.
- D. Compressive garments usually control discomfort in venous malformations, but usually exacerbate discomfort in GVM's.
- E. all of the above.

Correct Answer: Answer is E

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6. Regarding Gorham Stout Syndrome:

- A. It is usually a multisystem disease
- B. An effective treatment now exists
- C. It is predominantly a venous malformation
- D. There is usually a large soft tissue component.
- E. It is a lymphatic malformation which principally involves bone.

Correct Answer: Answer is E Gorham Stout malformation principally involves bone and, to a lesser extent, adjacent soft tissues. Any soft tissue component is minor. The defining characteristic is

disappearance of bone rather than a soft tissue mass in bone. It predominantly involves the upper limb and shoulder girdle.

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7. Regarding the use of Bleomycin, which of the following statements are true:

- A. It is FDA approved for intrapleural administration to control malignant pleural effusions.
- B. It produces markedly less swelling when treating venous malformations than ethanol and detergents
- C. In patients where the cumulative dose does not exceed 300 units, no documented case of pulmonary fibrosis has been reported.
- D. Bleomycin has been found to slightly less effective than ethanol in treating venous malformations of the head and neck, but to have significantly fewer complications.
- E. All of the above

Correct Answer: E. Although useful for treating venous malformations where swelling must be minimized, large confluent venous malformations do not respond well to Bleomycin.

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8. Regarding Doxycycline as a sclerosant agent:

- A. It is generally injected as a solution of 10 mgs per ml, in volumes up to 100mls.
- B. It commonly causes discoloration of developing dentition in children under 5 years
- C. Hyperglycemia is a recognized complication in neonates
- D. Injection is generally painless
- E. Nerve injury, although rare, is generally permanent.

Correct Answer: A.

References

1. Discoloration of teeth, in doses currently commonly used, is minimal or nonexistent. Hypoglycemia has been reported as a rare complication. Injection is painful, although probably less so than ethanol. Nerve injury has been reported in 14% of patients, but usually resolves. *J Pediatr Surg.* 2011 Nov;46(11):2083-95. doi: 10.1016/j.jpedsurg.2011.07.004.

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 4. Percutaneous sclerotherapy for lymphatic malformations: a retrospective analysis of patient-evaluated improvement. Alomari AI¹, Karian VE, Lord DJ, Padua HM, Burrows PE.
- 9. Austrian dermatologist Frederick Parkes Weber (1863-1962) is immortalized in which of the following conditions:**
- A. Parkes Weber syndrome
 - B. Sturge Weber syndrome
 - C. Rendu-Osler-Weber syndrome
 - D. Weber-Christian syndrome
 - E. All of the above.

Answer: E. His name is attached to six syndromes, three of which are composed of vascular anomalies.

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Treatment of Weber-Christian Disease R. Benson and P. D. Fowler

New Developments in Renovascular Hypertension

Derek J. Roebuck, MD

- 10. Which of the following endovascular techniques is not applicable to the treatment of renovascular hypertension in children?**
- A. Angioplasty
 - B. Stent insertion
 - C. Aneurysm coiling
 - D. Renal denervation
 - E. Ethanol ablation

Correct Answer: D

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Rationales

Although renal denervation using endovascular devices has been enthusiastically adopted in some countries, these devices do not have FDA approval, and the results of a recent randomized controlled trial with a sham operation arm suggest that the technique does not produce a

clinically-relevant reduction in blood pressure. All of the other techniques are applicable in appropriately selected patients. The results of angioplasty have been reported from numerous centers (1-5). The use of stents has been less well covered in the literature. There are several case reports, but no published long-term follow-up. Medium-term follow-up (5 years) in a group of 15 children has been presented in abstract form (6), but not in a peer-reviewed publication. Renal artery aneurysms may cause hypertension in childhood (7). Coiling or other endovascular treatments may have a role in their treatment. Ethanol ablation is probably the best treatment for stenosis or occlusion of small intrarenal arteries (8).

11. Which of the following is not an indication for insertion of a covered or uncovered stent in the renal artery of a child?

- A. Failure to abolish the waist of the angioplasty balloon with inflation at maximum rated ("burst") pressure
- B. Arterial rupture following angioplasty that does not respond to repeated reinflation of an angioplasty balloon
- C. Flow-limiting dissection following angioplasty that does not respond to repeated reinflation of an angioplasty balloon
- D. Immediate rethrombosis following recanalization of an occluded renal artery
- E. Repeated restenosis following technically and clinically successful angioplasties

Correct Answer: A

Rationale

This is a contraindication to stent insertion, because it will not be possible to deploy the stent at its maximum diameter. A better alternative would be to try a cutting balloon (9). The other options are all potential or definite indications for stenting (10). Repeated restenosis can be treated by repeated angioplasty, but in some patients stenting may be a reasonable alternative (11). Firth Cozens, J. and Greenhalgh J. (1997). "Doctors' perceptions of the links between stress and lowered clinical care." *Social Science & Medicine* 44(7): 1017-1022.

12. Which of the following non-invasive imaging tests has a sensitivity of >95% in the detection of a renovascular cause for pediatric hypertension?

- A. Ultrasound with pulsed-wave and color Doppler studies
- B. 256 detector row computed tomography angiography
- C. Magnetic resonance angiography
- D. Angiotensin converting enzyme inhibitor primed scintigraphy
- E. None of the above

Correct Answer: E

Rationale

A significant proportion (possibly as high as 50%) of renovascular lesions in children with hypertension involve small intrarenal branches of the renal artery or accessory arteries (12). Despite significant advances in technology (13), no non-invasive imaging technique has yet been shown to detect these with sufficient sensitivity to avoid the need for catheter angiography (14, 15).

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Venous Malformations: How to Stay Out of Trouble and Not Undertreat

Patricia E. Burrows, MD

13. **A superficial VM of the elbow area appears hypervascular on MRA and ultrasonography. Which of the following is the best treatment approach?**
 - A. Perform angiography and embolize the feeding arteries with liquid embolic agent.
 - B. Perform angiography, embolize the feeding arteries with particles and sclerose the VM percutaneously.
 - C. Perform angiography and sclerose the VM percutaneously.
 - D. Percutaneously sclerose the VM.
 - E. Do nothing because the lesion will involute spontaneously.

Correct Answer: D

Rationale

Answer is D. Many venous malformations are supplied by tiny muscular cutaneous arteries that may be exaggerated on MR angiography and color Doppler ultrasonography. Diagnostic catheter angiography usually confirms these mildly dilated arteries, which results in opacification of tiny peripheral components of the VM in the early venous phase. The bulk of the channels making up the VM are not opacified by arterial contrast injection. Arterial embolization does not result in thrombosis or shrinkage of the VM and is therefore ineffective in treatment of this anomaly. Appropriate treatment involves direct percutaneous cannulation of the venous malformation, confirmation of injection site by contrast injection, and injection of a sclerosing agent.

- A. Is not correct. The dilated arteries that supply a VM are also nutritive arteries to the skin and muscles and nerves in their distribution. Therefore, embolization of these vessels with liquid embolic agents typically results in severe tissue necrosis, often including peripheral nerves.
- B. Is not correct. While not as risky as A., embolization of the nutritive arteries with particles is ineffective in the treatment of VM and is thought to increase the risk of tissue necrosis with sclerotherapy.
- C. Is not correct. Angiography is rarely necessary to diagnose a VM.
- E. Is not correct. Venous malformations do not involute spontaneously.

14. A patient with diffuse VM has a fibrinogen level of 75 and clinical indications for sclerotherapy. Which of the following is best?

- A. Avoid anticoagulation because it may cause bleeding during sclerotherapy.
- B. Anticoagulate the patient for 24 hours after the procedure .
- C. Anticoagulate the patient for 2 weeks before and 2 weeks after the procedure.
- D. Prescribe aspirin for two weeks prior to sclerotherapy.
- E. Perform sclerotherapy and send the patient to the OR for resection the following day.

Correct Answer: C

Rationale

Answer is C. Patients with extensive VM often have intralesional consumption due to ongoing thrombosis, resulting in depression of fibrinogen levels and elevation of PT and INR. Administration of low molecular weight heparin [prophylactic dose] typically suppresses intralesional thrombosis, resulting in elevation of fibrinogen levels and normalization of INR. One to two weeks appears to be an adequate length of time for this affect. Continued anticoagulation after the procedure is recommended to minimize thrombosis and worsened coagulopathy.

- A. Is not correct. Unless the patient is actively bleeding, anticoagulation will actually decrease the risk of bleeding, because it will decrease consumption of clotting proteins.
- B. Is not correct. The patient should be anticoagulated before the procedure to improve coagulation status making the patient more receptive to sclerotherapy and less likely to develop severe coagulopathy after sclerotherapy.
- C. Is not correct. Aspirin may be mildly effective in decreasing intralesional clotting, but efficacy in terms of restoring fibrinogen levels has not been shown.
- D. Is not correct. Coagulopathy usually worsens after sclerotherapy if the patient hasn't been prepared by anticoagulation. Severe bleeding can result from open surgery.

15. Which of the following techniques is most likely to cause acute drop in oxygen saturation during sclerotherapy?
- A. Sclerotherapy of a diffuse VM with foam.
 - B. Sclerotherapy of a focal VM with ethanol
 - C. Sclerotherapy of a focal VM with foam without outflow occlusion
 - D. Sclerotherapy of a VM of conducting veins (type D) with ethanol using an automated tourniquet system to occlude flow for 10 minutes.
 - E. Sclerotherapy of VM of conducting veins with foam after occluding the outflow with coils or endovenous laser therapy

Correct Answer: D

Rationale

Answer is D. Ethanol sclerotherapy is known to cause acute desaturation as well as cardiomechanical dissociation with cardiac arrest when injected into non-sequestered VMs. Mechanisms could involve diffuse micro-embolism or pulmonary arterial vasospasm. It is recommended that individual aliquots of ethanol be limited to 0.1 ML per kilogram, with at least 10 min. between injections. However, incidents have occurred with smaller volumes. Injection of a large volume of ethanol into a non-sequestered VM behind a tourniquet is not recommended, because on release of the tourniquet, the ethanol associated with partly clotted veins is released acutely into the circulation.

- A. Is not correct. Release of foam into the systemic circulation is less likely to cause desaturation than ethanol. This technique can result in deep vein thrombosis, pulmonary emboli and early recanalization.
- B. Is not correct. Most focal VMs have minimal drainage to the conducting veins.
- C. Is not correct. In treating focal VM with foam, very little sclerosant
- E. Is not correct. Injection of any sclerosant into a sequestered venous malformation is unlikely to cause hypoxia. The same is true after permanent outflow occlusion.