ADULT CONGENITAL HEART DISEASE

1. Echo (TTE) allows assessment of **ASD** location, size, & associated manifestations e.g. right heart size, function & pulmonary pressure.
2. TEE is recommended if TTE is suboptimal, associated abnormalities are suspected, and is often required for diagnosis in sinus venosus.
3. Ostium secundum is most common **ASD** type (75% of cases), usually isolated congenital anomaly. L-R shunt causes right-sided dilatation.
4. Ostium primum **ASD** is usually associated with cleft anterior mitral or tricuspid valve leaflets (causing MR or TR), and VSD.
5. Sinus venosus **ASD** is usually associated with anomalous right-sided pulmonary veins connecting to the right atrium or SVC.
6. **ASD** should be considered in patients with new atrial arrhythmias.
7. Paradoxical embolism is a risk for all **ASD** patients regardless of size.
8. **ASDs** manifest fixed splitting of S2 and absence of sinus arrhythmia.
9. **ASD** is closed if there is right-sided volume overload or symptoms.
10. Most patients with **PFO** are asymptomatic. **PFO** closure may be considered in recurrent cryptogenic stroke despite medical therapy.
11. P*erimembranous* **VSDs**, most common, 80% in adults.
12. S*ubpulm./outlet,* 6% non-Asians, 33% Asians, rare spont. closure.
13. M*uscular,* common spont. closure, 20% in infants, lower in adults.
14. I*nlet,* rare in adults; Down syndrome in AV septal defect complex.
15. Small **VSDs** cause small shunts and loud holosystolic murmur; no LV volume overload and no pulmonary artery hypertension.
16. Moderate-sized **VSDs** and small to moderate L-R shunts cause LV volume overload and pulmonary artery hypertension with mild CHF.
17. Large **VSDs** are detected in childhood; without early closure, cause irreversible pulm. HTN with R – L shunt (Eisenmenger syndrome).
18. TTE allows identification of **VSD**, number & location; PA pressures, AI, TR. MRI is useful in inlet or apical **VSDs**, not seen by TTE.
19. Children and adults with isolated small **VSDs** rarely require closure.
20. **VSD** closure is recommended in adults with progressive AI or TR, progressive LV volume overload, and, recurrent endocarditis.
21. Unoperated aortic **coarctation** has 75% mortality by 46 yrs of age.
22. **Coarctation** is diagnosed by HTN in upper extremities & lower BP in lower extremities, resulting in radial – femoral pulse delay.
23. TTE confirms diagnsosis of **coarctation** and associated bicuspid AV (50% of cases). CMR & CT are preferred for imaging the aorta.
24. Auscultatory findings in **coarctation** include a systolic murmur & S4.
25. Surgical or percutaneous treatment of **coarctation** is recommended when the systolic gradient across the coarct is > 30 mmHg.
26. **PDA** is associated with maternal rubella & prematurity.
27. Adults with a moderate L – R **PDA** shunt present with left-sided cardiac enlargement, CHF symptoms and a machinery-type murmur.
28. Adults with previously unrecognized **PDA** may present with cyanosis and clubbing affecting the feet more than the hands.
29. TTE with Color Doppler is usually diagnostic of **PDA**; if nondiagnostic, cath helps identify shunting, PVR, and reactivity of the vascular bed.
30. Transcatheter/surgical closure in moderate-size **PDA** or endocarditis, without irreversible pulm. HTN. Otherwise, observation.

31. Valvular pulmonary stenosis is usually an isolated congenital abnormality; it may be associated with Noonan syndrome.
32. Severe pulm. stenosis may be tolerated for yrs without symptoms.
33. TTE & Doppler confirms the diagnosis and determines the severity of pulmonary stenosis and the degree of right ventricular hypertrophy.
34. Intervention is recommended when the peak systolic gradient is > 50 mmHg, when RVH is present, or when the patient is symptomatic.
35. Preferred pulmonic stenosis therapy is balloon valvuloplasty. cardiac surveillance is needed lifelong post valvotomy or replacement.
36. Post-**ASD**-repair: afib incidence increases with age at repair, device dislocation causing arrhythmia, peric. effusion, aortic erosion & SCD.
37. Post-**VSD**-repair: residual/recurrent **VSD**, AI, TR, endocarditis, heart block, arrhythmia & LV dysfunction. Need for reop. is uncommon.
38. Post-**coarct**-repair: persistent/recurrent HTN (50 – 75%), CAD, aortic aneurysm or dissection, endocarditis, AV disease, recoarct; may require reop. Age at initial repair is predictive of long-term survival.
39. **Tetralogy of Fallot** is most common cyanotic congenital heart disease; unoperated adults are rare. 35-yr survival postop is 85%.
40. Post-**TOF**-repair: PR (at RVOT annular patch); enlarged RV, TR, A/V arrhythmias; residual RVOT obstruction, PS, or VSD; RV or LV systolic & diastolic dysfunction, VT & SCD. Reop is commonly required.
41. **Cyanotic** congenital heart disease disposes to hemostatic problems, scoliosis, painful arthropathy/arthritis, gallstones, pulm hemorrhage or thrombus, paradoxical cerebral emboli/abscess, renal disease.
42. **Cyanotic** patients have compensated erythrocytosis with stable hemoglobins & require no intervention.
43. **Cyanotic** patients who have hyperviscosity (headaches and reduced concentration), dehydration should be excluded before phlebotomy.
44. **Cyanotic** patients are at risk of paradoxical embolism: from IV line air- air filters are used; from DVT- early ambulation/SCDs are used. Anticoagulation is avoided (platelet dysfunction/bleeding tendency).
45. Pregnancy has 50% maternal mortality in **cyanotics** & Eisenmenger.
46. Treatment options for symptomatic **Eisenmenger** syndrome patients include pulmonary vasodilators and lung or heart-lung transplant.
47. **Cyanotic/Eisenmenger** patients should be seen by adult congenital cardiac specialist annually. When hospitalized for medical or surgical problems, they should be seen & followed by such specialist.