PART ONE

Disease
ADRENAL & PITUITARY DISEASE

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OVERVIEW
- Pituitary adenomas represent 10–15% of intracranial neoplasms.
- Prevalence is 200/1,000,000; post-mortem incidence is 10–27%.
- Significant number are asymptomatic.
- Most pituitary tumors arise from anterior lobe (adenohypophysis)
  ➢ Majority are benign.
- 75% of tumors secrete hormones inappropriately
  ➢ Prolactin
  ➢ Growth hormone (GH): acromegaly
  ➢ Premature puberty or resumption of menstrual bleeding in postmenopausal women
  ➢ Adrenocorticotrophic hormone (ACTH): Cushing's syndrome
- Presentation variable
  ➢ Hormonal hypersecretion syndrome (see above)
  ➢ Mass effect (non-secreting macroadenomas >1 cm): visual disturbance (bitemporal hemianopsia/third nerve palsy) or increased ICP
  ➢ Nonspecific symptoms, such as headache, infertility, pituitary hypofunction or epilepsy (compression from non-functioning adenoma, secondary hyperprolactinemia, apoplexy or infarction)
  ➢ Incidental finding

FLUID & ELECTROLYTES
- Acromegaly
  ➢ Diabetes
### Adrenal & Pituitary Disease

- **Cushing's syndrome**
  - Diabetes
  - Hypernatremia, hypokalemia & alkalosis
  - Renal calculi

- **Hypopituitarism**
  - Hyponatremia, hyperkalemia & acidosis

### Cardiopulmonary

- **Acromegaly**
  - Hypertension
  - Cardiomegaly, impaired LV function (interstitial fibrosis)
  - Obstructive sleep apnea (OSA): macroglossia, thickened pharyngeal tissues

- **Cushing's syndrome**
  - Hypertension
  - ECG: LVH, high-voltage QRS, inverted T waves; reversible w/ pituitary removal

### Hematologic

N/A

### Metabolic-Nutritional

- 5 distinct cell types plus null (functionally inert) cells
  - 50% somatotrophs: secrete GH
  - 10–25% lactotrophs: secrete prolactin
  - 15% corticotrophs: secrete ACTH
  - 10% gonadotrophs: secrete follicle-stimulating hormone (FSH) & luteinizing hormone (LH)
  - 5–10% thyrotrophs: secrete thyroid-stimulating hormone

- Production & release of anterior pituitary (adenohypophysis) hormones under control of hypothalamus:
  - Prolactin stimulated by prolactin-releasing hormone (PRH) & inhibited by dopamine
  - GH stimulated by growth hormone-releasing hormone & inhibited by somatostatin
Adrenal & Pituitary Disease  |  Adult Congenital Heart Disease

- Thyroid-stimulating hormone (TSH) stimulated by thyroid-releasing hormone (TRH)
- ACTH stimulated by corticotrophin-releasing hormone (CRH)

Anterior pituitary hormones are under feedback control.
- Insulin-like growth factor I directly inhibits GH but also stimulates secretion of somatostatin.
- Thyroid hormones inhibit both TSH & TRH.
- FSH & LH have both inhibitory & stimulatory effects on the pituitary & hypothalamus.

Posterior (neurohypophysis) secretes oxytocin & arginine vasopressin (AVP).

**GASTROINTESTINAL**
- Cushing’s syndrome is associated w/ gastroesophageal reflux disease (GERD).

**NEUROPSYCHIATRIC**
- Cushing’s syndrome is associated w/ mental disturbance.

**ADULT CONGENITAL HEART DISEASE**

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**OVERVIEW**
- Adults w/ CHD may remain undiagnosed until adulthood.
- Adults may have had 1 or more corrective or palliative cardiac procedures.
- Clinical picture: ranges from essentially normal physiology to critically ill
- End-stage adult CHD: severe pulmonary hypertension (PHT) & Eisenmenger physiology

Congenital defects fall into 3 groups:
- Shunt lesions resulting in increased pulmonary blood flow (PBF)
Adult Congenital Heart Disease

- Shunt lesions resulting in decreased PBF
- Obstructive lesions, resulting in increased cardiac work & impaired blood flow distal to the obstruction

Two major consequences of the abnormal anatomy:
- Cyanosis: decreased pulmonary artery (PA) blood flow or mixing of systemic & pulmonary venous (PV) return
- CHF: increased PA blood flow, from excessive PV return, or impaired PV return from obstruction or impaired forward cardiac output (CO). Both lead to pulmonary vascular congestion.

Classification of CHD also depends on surgical status:
- Anatomy may be unchanged or previously surgically altered by a corrective or palliative procedure.
- Surgical correction of complex lesions (eg, single ventricles) is often staged.
- Corrective procedures can themselves induce heart block or impair ventricular function.

Acyanotic lesions
- Ventricular septal defect (VSD)
  - Chronic volume overload results in abnormal ventricular function. Pts repaired after 5 yrs of age are likely to have residual ventricular dysfunction.
  - Nonrestrictive or moderately restrictive VSDs cause pulmonary vascular pressure/volume overload & PHT. Ultimately, reversal of shunt results in Eisenmenger physiology.
- Atrial septal defect (ASD)
  - Simple ostium secundum defect is the most common shunt lesion in adults.
  - May be asymptomatic & permit survival into late adulthood
Adult Congenital Heart Disease

➤ Most pts >60 yrs old are symptomatic: increased systemic arterial pressure & decreased ventricular compliance cause increased L-to-R shunt through the ASD.
➤ Atrial tachyarrhythmias are common > 40 yrs.
➤ PHT develops > 40 yrs.
➤ Pts w/ normal pulmonary pressures corrected surgically before 24 yrs have normal survival.
➤ Paradoxical emboli (from atrium to cerebral vessels) may occur because of preferential streaming of blood flow from IVC across foramen ovale (secundum ASD), especially during sudden increases in PVR (coughing, etc.).

■ Partial anomalous pulmonary venous connection
  ➤ Partial pulmonary venous return to right atrium (RA)
  ➤ Wide variety of anomalous pulmonary venous connections, usually w/ sinus venosus ASD
  ➤ Physiology & natural history similar to ASD for similar degree of L-to-R shunt
  ➤ SA node dysfunction possible if sinus venosus type
  ➤ Scimitar syndrome: anomalous connection of all right PV to IVC, associated w/ hypoplastic right lung (arterial circulation to right lung from descending thoracic aorta)

■ Coarctation of the aorta
  ➤ Commonly associated w/ bicuspid aortic valve
  ➤ Severe coarctation presents w/ heart failure within first wks of life.
  ➤ Milder coarctation: survive into adulthood
  ➤ Later repairs associated w/:
    • High incidence of hypertension in pts repaired late
    • Increased incidence of premature coronary artery disease

■ Pulmonary valve stenosis
  ➤ Most stenosis is noncritical; survival into adulthood is the rule.
Adult Congenital Heart Disease

- Associated w/ right ventricular (RV) hypertrophy & eventual RV failure
- Commonly repaired w/ balloon valvuloplasty

- Congenitally corrected transposition of the great arteries (L-transposition, ventricular inversion)
  - Blood flow: RA \(\rightarrow\) MV \(\rightarrow\) LV \(\rightarrow\) PA \(\rightarrow\) lungs \(\rightarrow\) LA \(\rightarrow\) tricuspid valve \(\rightarrow\) RV \(\rightarrow\) aorta
  - Aorta located anterior & to the left of the main PA
  - Anatomic RV serves as systemic ventricle.
  - RV eventually vulnerable to failure
  - High incidence of heart block

- Ebstein’s anomaly of the tricuspid valve
  - Abnormal apical displacement of the tricuspid tissue \(\rightarrow\) atrialized RV w/ tricuspid regurgitation
  - Sometimes w/ pulmonic stenosis

Cyanotic lesions

- Tetralogy of Fallot (TOF)
  - Pulmonic stenosis, VSD, overriding aorta, RVH
  - Most common cyanotic lesion encountered in older pts
  - Uncorrected: 25% survival to adolescence; 3% >40 yrs
  - After early repair, LV ok, RV usually impaired
  - “Tet spell”: increased R-to-L shunt secondary to decreased SVR; treated w/ phenylephrine to increase SVR (pulmonary resistance “fixed” from stenotic pulmonic valve)
  - “Tet spells” rare in older pts w/ uncorrected TOF
  - After repair, right bundle branch block (RBBB) common, sometimes associated w/ left anterior hemiblock

- Palliated tetralogy of Fallot
  - Palliative shunts (Blalock-Taussig, Waterston-Cooley, Potts or Gore-Tex central aortopulmonary) to increase PBF
  - PBF dependent on relative PVR vs. SVR

- Complete transposition of the great arteries (D-transposition)
  - Incompatible w/ life unless adequate mixing
4 types of repair

- Mustard or Senning (atrial repair): systemic venous return → LV → PA & pulmonary venous return → RV → aorta
- Arterial switch: aorta & coronary arteries transposed to arise from LV & PA moved to arise from RV; most common repair now
- Rastelli: LV outflow channeled through the VSD to aorta & RV connected to PA by external conduit
- Palliative atrial repair in presence of a VSD & pulmonic valve disease (least common)

Post-repair physiology depends on repair type:

- An RV that must function as a systemic ventricle is vulnerable to failure.
- Atrial repair is associated with high incidence of electrophysiologic problems (SA node dysfunction, AV block, SVT, etc.).

Single ventricle physiology

- Include those with tricuspid atresia or a single, double inlet ventricle, usually a morphologic left ventricle
- Survival to adulthood depends on the anatomy: pulmonary stenosis (protects pulmonary vascular bed) & competent atroventricular valve(s) increase survival.
- Generally pts with single LV do better than pts with single RV (morphologic).
- Pts with unrestricted PBF typically develop pulmonary vascular disease.
- Pts with restricted PBF may have required palliative shunts to increase PBF (to treat hypoxia).
- Aortopulmonary shunts result in volume overload of the single ventricle; over time this results in ventricular failure.
- Cavopulmonary shunts (Glenn) increase PBF without volume overload of the single ventricle.
Adult Congenital Heart Disease

Fontan procedure
- RA to PA connection for “correction” of single-ventricle hearts
- Two-stage procedure
  - Step 1: anastomosis of SVC to undivided right PA = bidirectional Glenn shunt
  - Step 2: IVC connected to PA via Gore-Tex graft through RA
- Graft usually baffled into LA so that some R-to-L shunt persists as safety valve in cases of high pulmonary resistance
  - Fluid retention (effusions, ascites)
  - Forward flow depends on CVP exceeding LAP to provide driving force for PA blood flow.
  - Must have adequate intravascular volume & avoid increased PVR (see above)

Truncus arteriosus
- Truncus arteriosus w/ unobstructed PA origin from the ascending aorta rapidly results in fatal CHF w/out correction.
- Survival is permitted by pulmonary vascular inflow stenosis/disease sufficient to limit L-to-R shunt (but cyanosis is increased).
- Truncal valve often malformed & incompetent; regurgitation is biventricular & can be severe
- Adult survivors: repair by closure of VSD & interposition of valved conduit between RV & PA
  - The truncal valve remains as the aortic valve & incompetence should be treated like aortic insufficiency.
  - The valved RV to PA conduit is prone to stenosis & insufficiency RV failure.
  - The conduit lies just beneath the sternum & is vulnerable to sternotomy.

Total anomalous pulmonary venous connection
- Obligatory R-to-L shunt resulting from pulmonary veins emptying into RA
Adult Congenital Heart Disease

Pts w/ supradiaphragmatic PV connection, low PVR & a large ASD have natural history similar to that of isolated nonrestrictive ASD but w/ systemic arterial saturations in the low 90s.

Eisenmenger physiology/pulmonary vascular disease

- PVR at or above SVR w/ resultant R-to-L shunt
- Anatomy: nonrestrictive communications at great arterial, ventricular or atrial level, by heart w/ single ventricle or two ventricles w/ or w/out ventricular inversion
- Excessive PBF & pressure result in changes in PA vasculature & ultimately fixed high PVR.
- Confers high perioperative risk:
  - Represents most of unoperated adults w/ CHD who are referred for preop anesthetic evaluation
  - Fixed PVR precludes rapid adaptation to intraoperative hemodynamic changes.
  - Variations in SVR produce changes in the magnitude of R-to-L shunting (decreased SVR leads to increased R-to-L shunt).
  - Hypovolemia, hypotension, vasodilators & regional anesthesia are poorly tolerated & exacerbate R-to-L shunt.
  - Distorted cardiac anatomy & R-to-L shunt obviate PA catheters (hazardous placement, inaccurate CO). Relative resistance of PVR vs. SVR can be monitored by changes in systemic arterial pulse oximetry (SpO₂).
  - Fixed high PVR is unresponsive to pharmacologic treatment but exacerbated by cold, acidosis, hypercarbia, hypoxia, endogenous or exogenous catecholamines w/ alpha-adrenergic effects.

Aortopulmonary shunts

- Used in any defect associated w/ low PBF
- May be palliative until reparative procedure or definitive procedure can be done