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Reexpansion Pulmonary Edema

Reinflation of a collapsed lung in a few cases can lead to pulmonary edema of the reexpanded lung. This complication termed reexpansion pulmonary edema (RPE) may occur after treatment of a lung that has collapsed after pneumothorax, pleural effusion or thoracoscopic resection of a mediastinal tumor in less than 1% of all cases. The clinical presentation of RPE is characterized by a rapid onset of dyspnea and tachypnea with symptoms developing upon one hour of reexpansion of the lung. Simple chest films may reveal interstitial opacities, consolidations with air bronchograms, and fissural inflammation. Risk factors for RPE includes the degree and chronicity of lung collapse (usually greater than 72 hours), great amount of pleural air or fluid, high speed of reexpansion, use of high negative pressure to do so, hypertension, hypoxemia and previous lung disease. Severe RPE can lead to bradycardia, hypotension, cardiopulmonary arrest, and death. RPE occurs most frequently in a chronically collapsed lung, which is then rapidly reinflated using high suction. The endpoint of the reexpansion injury is an increase in permeability of the endovascular cells and increase in hydrostatic pressure, which then leads to the pulmonary edema. Both of them cause fluid and protein overflow into the pulmonary interstitial space and alveoli, leading to pulmonary edema As a form of prevention the use of immediate suction to a chest tube placed for reexpanding a collapse lung after pneumothorax or effusion should be avoided as this is a precipitant factor for development of RPE. It is preferably that the lung reexpands more gradually. Treatment once RPE occurs remains supportive. The cornerstone is positive-pressure mechanical ventilation and utilization of positive end-expiratory pressure (PEEP) to reexpand collapsed alveoli, increase functional residual capacity, and reduce shunting. Treatment also may include steroids, diuresis and vasopressor support. In the future, the use of agents such as monoclonal antibody to IL-8 or XOD antagonists may be useful for prevention or treatment of RPE.

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Turner Syndrome

Turner syndrome (TS), also known as gonadal dysgenesis, is a fairly common chromosomal abnormality occurring in females. It is characterized by short stature, web neck, cubitus valgus, low hairline, short 4th and 5th metacarpals, sexual infantilism, bilateral rudimentary streak gonads and primary amenorrhea among other defects. Fifty percent of TS have sex chromosome monosomy with a 45,X karyotype and the remaining patients have either mosaicism with a 45,X cell line or a structural X anomaly. Girls with typical TS with bilateral streak gonads are not at risk for development of gonadoblastoma in their dysgenetic gonads and gonadectomy is not indicated. In routine cytogenetic analysis the Y chromosome or Y-specific sequence is present in 5 to 10% of patient with TS. A higher incidence of Y-chromosome material has been reported when polymerase chain reaction (PCR) and fluorescent in situ hybridization (FISH) techniques are used in addition to peripheral blood karyotyping. Dysgenetic gonads with the presence of a Y chromosome or translocated fragment have a significant risk of developing germ cell tumors, specifically gonadoblastoma, in their dysgenetic gonads. The risk ranged between 25% and 75% and increases with advancing age. Gonadoblastoma is the most commonly found tumor and considered an in-situ neoplastic lesion with further risk for malignant transformation to a dysgerminoma or another invasive germ cell tumor (such as yolk sac tumor, embryonal cell carcinoma and malignant teratoma). Gonadoblastoma is mostly seen in those TS with a 45,X/46,XY karyotype. Prophylactic gonadectomy is currently recommended in all TS patients with Y-chromosome material. Laparoscopic salpingo-oophorectomy of the streak gonad is the preferred procedure. This approach eliminates the complication of future ectopic tubal pregnancy along with the possibility of tubal malignancy.

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Aortoesophageal Fistula

Aortoesophageal fistula (AEF) is a very rare, serious and almost lethal condition in a

child if not diagnosed promptly and managed expedite. Foreign body ingestion remains the commonest cause of AEF seen in children. AEF presents with the classic triad of midthoracic pain, a small sentinel hemorrhage followed later by an exsanguinating hemorrhage. Any child presenting with these symptoms should be suspected of having an AEF until proven otherwise. Most children presenting with an AEF will have had a congenital cardiac or vascular anomaly which required surgical correction or have ingested a foreign body such as fish bone, chicken bones or stuck button batteries. The hemodynamic stability of the patient dictates whether further diagnostic investigations are appropriate. Diagnosis can be established using esophagoscopy, CT-angio Scan, conventional arteriography or upper gastrointestinal contrast imaging. Temporary control of the bleeding from an AEF can be obtained using Sengstaken-Blakemore tube or bedside placement of an aortic occlusion balloon until either diagnosis or definitive surgery can be performed. Other temporizing measures to control hemorrhage include endovascular stents, hemoclips at endoscopy, and radiographic embolization. Surgical exploration of the thoracic esophagus and aorta with repair of the fistula, preferably under cardiopulmonary bypass, remains the only hope for cure and survival for children with AEF. In cases of stuck battery in the esophagus, they should undergo emergency endoscopic removal and inspection of the esophageal mucosa. Failure to remove batteries within two hours can lead to esophageal necrosis and aortoesophageal fistulas. The mortality of AEF is extremely high.

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