Respiratory

Management of anaesthesia for a teenager with cystic fibrosis

Smoking
Management of anaesthesia for a teenager with cystic fibrosis

Cystic fibrosis
AR inherited condition: 1/2500 Caucasians
Chromosome 7 mutation, multiple abnormalities described
Expressed as defective Cl channels
High Cl concentration in secretions, increased viscosity
Sweat Cl > 60 mmol/l
Resultant respiratory, pancreatic, biliary dysfunction

Clinical problems
Respiratory
Impaired clearance of secretions, mucociliary dysfunction
Chronic infection, bronchiectasis, dyspnoea, excessive sputum
Air-trapping due to mucous plugging: COAD-like picture
Nasal polyps, chronic sinusitis
Complications
Respiratory failure: cyanosis, CO$_2$ retention
Haemoptysis, pneumothorax, cor pulmonale

GIT
Pancreatic exocrine failure
Malabsorption, malnutrition without enzyme supplementation
Chronic pancreatitis, acute exacerbations
Secondary endocrine failure: diabetes mellitus
Bile secretion impaired
Fat and fat-soluble vitamin malabsorption
Later cirrhosis and portal hypertension
Neonatal meconium ileus

Psychological
Many admissions and procedures
Chronic illness, medicalization
Longer survival resulting in more adult presentations

Assessment
Routine, plus
History related to CF
Admissions, current therapy, respiratory disease, diabetic control
Previous anaesthesia
Examination
General appearance, respiratory, cardiac focus
Signs of respiratory failure, right heart failure
Investigation
CXR, RFT, ABG
FBE, U&E, LFT, glucose
Recent cultures of sputum

Optimize
Consult with treating physician to achieve best respiratory function
Bronchodilators, saline nebs, antibiotics, physiotherapy

Technique
Regional avoids the need for intubation and risk of worsening infection
If GA required by nature of surgery, regional analgesia may result in better respiratory function postoperatively
Airway
Humidified gases, nebulized saline may be of benefit
Frequent suctioning
Avoid nasal intubation
Ventilation
As for COAD: long I:E, minimize airway pressures, slow rate
Increased FiO₂
Vigilance for pneumothorax

Circulation
Consider invasive monitoring if cor pulmonale: arterial line ± PA catheter

Other considerations
Management of diabetes: fasting, glucose monitoring
Choice of drugs suitable for impaired hepatic function

Postoperative
Level of care determined by severity of disease and extent of surgery
Active physiotherapy and early mobilization
Smoking