

Anaesthesia recommendations for **Scimitar syndrome**

Disease name: Scimitar Syndrome

ICD 10: Q26.8

Synonyms: Congenital Pulmonary Venolobar Syndrome, Pulmonary Venous Return Anomaly, Partial/Total Anomalous Pulmonary Venous Return (P/TAPVR)

Disease summary: Scimitar syndrome (SS) is a rare congenital heart defect (CHD) with a frequency of 1-3 cases per 100.000 live births and a suspected autosomal inheritance. It results in a partial to total anomalous venous drainage of the right lung into the inferior vena cava (IVC). SS may be associated with atrial septum defects, aberrant systemic arterial supply (SAS) to the right lung through aorto-pulmonary collaterals and other heart defects. Anatomical sequelae include right lung hypoplasia and left to right shunting. This can lead to recurring lower respiratory tract infections (LRTI) and right ventricular volume overload causing pulmonary artery hypertension and right ventricular failure. Clinical presentation can be highly variable. Severe forms present as cyanosis and acute heart failure of the new-born or infant. Dyspnoea on exertion is a possible common symptom. Mild forms may be incidental findings in asymptomatic adults. Diagnosis can be confirmed by echocardiogram or computed tomography showing the anomalous venous return. Treatment depends on symptom severity and may include correction of venous return in cases of severe heart failure. Patients with mild symptoms will usually be handled conservatively with diuretics and standard chronic heart failure medication.

Medicine is in constant progress; new clinical knowledge may not be in this guideline.



Recommendations are not rules or laws; they are a framework for clinical decision-making.

Every patient is unique; individual circumstances must guide clinical care.

The diagnosis may be wrong; if questionable, the diagnosis should be confirmed.



Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

Typical surgery

Surgical correction of anomalous venous return. Ligation of aorto-pulmonary collaterals in SAS. Closure of ASD. Surgical revision of scimitar vein conduits may be necessary. Lobectomy/ Pneumonectomy in lung hypoplasia and recurring LRTIs.

Surgery unrelated to SS.

Type of anaesthesia

There is no definite recommendation for or against either general or regional anaesthesia. There are reports of spinal, epidural and caudal anaesthesia without any complications. Circulatory effects of each technique must be anticipated and counter-balanced.

Intraoperative and perioperative factors that result in increases of pulmonary vascular resistance such as hypoxia, hypercarbia, hypothermia, acidosis, atelectasis, high airway pressure should be avoided.

Necessary additional pre-operative testing (beside standard care)

Effects on the cardio-circulatory and pulmonary system are highly variable and need to be investigated thoroughly in an elective setting.

Cardiac testing should include electrocardiography and echocardiography to assess right (and left) ventricular function, shunting fraction and pulmonary artery pressures.

Spirometry and blood gas analysis are helpful to evaluate pulmonary involvement. Plain chest X-ray may reveal the pathognomonic opacification caused by the scimitar vein.

Standard lab-work including liver-function tests (LFTs) are advisable.

Consulting a cardiologist with experience in congenital heart defects can be useful.

Particular preparation for airway management

No evidence of association of difficult airway and SS.

Particular preparation for transfusion or administration of blood products

Transfusion requirements will result from the nature of the surgical procedure. SS is not directly associated with bleeding disorders, but patients may be receiving anti-coagulants for associated heart defects.

Particular preparation for anticoagulation

Literature lacks evidence to guide anticoagulation in SS in general. Impaired mobility of severely affected patients may suggest a higher risk of perioperative thrombosis.

Particular precautions for positioning, transportation and mobilisation

Not reported.

Interactions of chronic disease and anaesthesia medications

Severe hypotension when nitrous oxide is combined with long term PDE-inhibitors such as sildenafil et al.

Anaesthetic procedure

Induction and maintenance of anaesthesia can be achieved by inhalational and/or intravenous techniques. Successful regional and neuro-axial techniques have been reported.

Adequate right-ventricular preload is a prerequisite. Factors that increase pulmonary vascular resistance such as hypoxia, hypercarbia, hypothermia, acidosis, atelectasis, high airway pressure must be avoided. High concentrations of volatile agents may impair hypoxic pulmonary vasoconstriction. A high-flow left-right shunt can increase hysteresis (i.e. administration-till-effect time interval). Avoid any air in IV lines and ports especially in presence of ASD. Bubble-catch filters are recommended. Consider antibiotic endocarditis prophylaxis.

Sufficient analgesia prevents sympathoadrenergic reactions and decreases right ventricular strain.

Particular or additional monitoring

In major surgery or advanced disease arterial cannulation for invasive blood pressure measurement and central venous line placement is recommended. Transoesophageal echocardiography and/or a pulmonary artery catheter may be considered in high risk scenarios. However, there is insufficient data to provide an evidence based recommendation for these invasive procedures.

Simultaneous pre- and post- ductal oxygen-saturation monitoring is advisable in neonates.

Possible complications

Increases in pulmonary vascular resistance leading to acute right ventricular failure. Cardiac arrhythmias. Lower respiratory tract infections. Respiratory failure.

Post-operative care

Postoperative care on ICU is recommended in all cases of major surgery and advanced disease. A high degree of suspicion is advisable in all other cases.

Disease-related acute problems and effect on anaesthesia and recovery

Ambulatory anaesthesia

Should only be considered on an individual basis in patients with mild disease and minor surgery.

Obstetrical anaesthesia

The additional circulatory burden in advanced pregnancy may result in volume overload and right ventricular failure even in previously asymptomatic patients. Miscarriages, prematurity and foetal growth-retardation are common and premature delivery may be necessary.

Literature suggests vaginal birth flanked by early and gradual epidural analgesia as based on expert opinion.

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Date last modified: **December 2018**

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Disclosure(s) The authors have no financial or other competing interest to disclose. This guideline was unfunded.

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Disclosure(s) The reviewer has no financial or other competing interest to disclose.
