Scoliosis is a lateral curvature and rotation of the thoraco-lumbar vertebrae with a resulting rib cage deformity. It may be idiopathic or secondary to neuromuscular disease, infection, tumour or injury (Table 1).

Curvature is measured using the Cobb angle (Fig. 1). A lateral curve of >10° is considered abnormal.1

Natural history
Adolescent idiopathic scoliosis is the most common form of scoliosis, and is most frequent in girls. In most cases the curvature remains small or may even resolve. However, if spinal curvature progresses, there is an increase in cosmetic deformity, back pain and chest cavity narrowing. More severe curves result in a restrictive lung defect and dyspnoea on exertion (>65°/C14). Uncorrected progression leads to respiratory failure, pulmonary hypertension and right heart failure (>100°/C14). Surgery is indicated once the curvature is >40°.

A restrictive lung defect attributable to scoliosis compounds the respiratory weakness associated with neuromuscular disease. Surgery may slow the decline in respiratory function and improve quality of life by improving posture and helping nursing care. Such patients should be offered stabilization before their cardio-respiratory dysfunction prevents surgery.

Surgical management
The aim of spinal deformity surgery is to correct the curve and fuse the spine, improving posture and halting the progression of pulmonary dysfunction. The approach may be posterior, anterior or combined depending on the cause and severity of the curvature. In the most commonly used posterior approach, the skin and supraspinous ligament are incised and paraspinal muscles reflected. The vertebral laminae are then decorticated, facet joints destroyed and spinous processes removed. Bone graft is packed over the raw decorticated surfaces and stainless steel rods (secured with pedicle screws or laminar hooks) are used to correct the deformity and provide stability for bony fusion.

The anterior approach involves a large thoraco-abdominal incision, exposure of the vertebral bodies and removal of the intervertebral discs to allow for greater movement. One lung ventilation is rarely necessary to

Table 1 Classification of scoliosis aetiology

<table>
<thead>
<tr>
<th>Category</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Idiopathic</td>
<td>Early onset (infantile)</td>
</tr>
<tr>
<td></td>
<td>Late onset (juvenile/adolescent)</td>
</tr>
<tr>
<td>Neuromuscular</td>
<td>Cerebral palsy</td>
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<tr>
<td></td>
<td>Myopathies</td>
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<tr>
<td></td>
<td>Polymyelitis</td>
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<tr>
<td></td>
<td>Syringomyelia</td>
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<tr>
<td></td>
<td>Friedreich’s ataxia</td>
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<tr>
<td>Congenital</td>
<td>Vertebral anomalies</td>
</tr>
<tr>
<td></td>
<td>Rib anomalies</td>
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<tr>
<td></td>
<td>Spinal dysraphism</td>
</tr>
<tr>
<td>Traumatic</td>
<td>Vertebral fractures</td>
</tr>
<tr>
<td></td>
<td>Radiation</td>
</tr>
<tr>
<td></td>
<td>Surgery</td>
</tr>
<tr>
<td>Syndromes</td>
<td>Marfan’s</td>
</tr>
<tr>
<td></td>
<td>Rheumatoid arthritis</td>
</tr>
<tr>
<td></td>
<td>Osteogenesis imperfecta</td>
</tr>
<tr>
<td></td>
<td>Mucopolysaccharide disorders</td>
</tr>
<tr>
<td></td>
<td>Neurofibromatosis</td>
</tr>
<tr>
<td>Neoplastic</td>
<td>Primary tumours</td>
</tr>
<tr>
<td></td>
<td>Secondary tumours</td>
</tr>
<tr>
<td>Infection</td>
<td>Tuberculosis</td>
</tr>
<tr>
<td></td>
<td>Osteomyelitis</td>
</tr>
</tbody>
</table>

Fig. 1 The Cobb Angle. Perpendicular lines are drawn from the outer surfaces of the upper and lower, maximally tilted, vertebrae. The angle formed by these intersecting perpendiculars determines the Cobb angle (X°).
improve surgical access, except in high thoracic curves. Combining
the anterior and posterior approaches in a single operation
results in a more rapid recovery and less time in hospital. In
our experience, there is no advantage in staging procedures
more than 1–2 weeks. The more high risk non-idiopathic patients
receive only posterior surgery because of the increased morbidity
with an anterior approach.

**Principles of anaesthetic management**

**Preoperative assessment**

The aetiology, location and degree of scoliosis should be noted.
All patients require a full history, physical examination and
appropriate investigations focusing on cardiovascular and
respiratory systems (Table 2).

**Idiopathic scoliosis**

In patients with idiopathic scoliosis a good exercise tolerance and
absence of respiratory symptoms indicates acceptable cardio-
respiratory reserve. Patients with more severe degrees of scoliosis
(>100°), right ventricular hypertrophy on electrocardiograph or
evidence of right heart failure on examination require echo-
cardiography. Spirometry is performed routinely on all patients
and will usually show a restrictive lung defect. There is evidence
that scoliosis surgery can be well tolerated despite severe restrictive
lung disease (FVC < 32%).3 Approximately 25% patients with
idiopathic scoliosis have mitral valve prolapse, but this is rarely
of clinical significance and antibiotic cover is given.

**Non-idiopathic scoliosis**

Preoperative assessment of patients with neuromuscular disease or
immobility is more difficult. They are neither able to give a history
of exercise tolerance nor perform spirometry adequately. Muscu-
lar dystrophies may be complicated by subclinical cardiomy-
opathy. More than 50% of patients with Duchenne muscular
dystrophy have some degree of dilated cardiomyopathy and
an ejection fraction <45% by 15 yr of age.4 Any reduction in
ejection fraction may mean difficulties coping with the rapid
fluid shifts during surgery. Echocardiography is required to assess
left ventricular function in these patients; however, a normal
study does not exclude significant pathology. Pharmacological
stress echocardiography may give a better indication of cardiac
function under anaesthesia but is less readily available.

The use of invasive monitoring lines and catheters along
with postoperative analgesia plan should be explained fully to
the patient and family. Sedative premedication with oral mid-
azolam (0.5 mg kg⁻¹) can be offered. Patients with Duchenne
muscular dystrophy may be on corticosteroid therapy and
require perioperative supplementation.

**Anaesthetic technique**

The aim is to maintain a stable anaesthetic depth allowing
for intraoperative neurophysiological monitoring (see below)
and this can be achieved using various anaesthetic techniques.
We use an i.v. induction of propofol followed by a non-
depolarizing neuromuscular blocking drug and tracheal intub-
ation with an armoured tracheal tube. Anaesthesia is maintained
by sevoflurane at 0.6 MAC in air and oxygen, with an infusion of
remifentanil. A bolus of i.v. morphine is given towards the end
of surgery. Succinylcholine is contraindicated in patients with
muscular dystrophy because of the risk of rhabdomyolysis,
hyperkalaemia and cardiac arrest.

**Positioning**

Turning the anaesthetized patient prone, often for long periods,
may cause a number of mechanical or physiological problems.
It requires good teamwork and attention to detail. Accidental
extubation or dislodgement of intravascular and urinary catheters
can occur, and monitoring during the turn is difficult. The require-
ments are an X-ray compatible table, a four-poster or Wilson
frame and full-length foam padding. These frames allow free
abdominal movement during respiration, helping ventilation by
improving compliance. They also prevent an increase in intra-
abdominal pressure and reduced venous return, which results in
epidural venous congestion and increased blood loss.

Once in the prone position, monitoring should be recom-
mended and the chest auscultated for bilateral air entry. The head
should be positioned carefully and secured to prevent excessive
flexion, extension or rotation of the neck. Postoperative blind-
ness has been attributed to external eye pressure reducing optic
nerve and retinal perfusion; therefore, the eyes should be pro-
tected and checked frequently throughout surgery. The arms
should not be placed >90° in abduction or forward flexion.
Vulnerable areas such as peripheral nerves and genitalia should
be protected from compression and soft tissue damage.

**Monitoring and temperature control**

Scoliosis surgery is associated with substantial blood and heat
loss, with the potential for haemodynamic instability. In addition
to standard paediatric general anaesthetic monitoring, invasive
arterial pressure monitoring and a urinary catheter are essential.
Two large peripheral i.v. cannulae are sited. In our unit, a central
venous cannula is placed if there is significant comorbidity

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**Table 2 Preoperative investigations for scoliosis surgery**

<table>
<thead>
<tr>
<th>Routine investigations</th>
<th>Additional investigations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Plain chest X-ray</td>
<td>Arterial blood gases— if</td>
</tr>
<tr>
<td>Pulmonary function tests—</td>
<td>spirometry not possible</td>
</tr>
<tr>
<td>FEV₁ and FVC</td>
<td>ECG and Echocardiography</td>
</tr>
<tr>
<td>Blood tests—</td>
<td>(non-idiopathic scoliosis)</td>
</tr>
<tr>
<td>Full blood count</td>
<td></td>
</tr>
<tr>
<td>Coagulation screen</td>
<td></td>
</tr>
<tr>
<td>Urea and electrolytes</td>
<td></td>
</tr>
<tr>
<td>Calcium and phosphate</td>
<td></td>
</tr>
<tr>
<td>Blood cross-match</td>
<td></td>
</tr>
</tbody>
</table>
scoliosis surgery can result in excessive blood loss which can exceed circulating blood volume. Coagulopathy (dilutional and consumptive) can develop and prevent haemostasis. Factors influencing the degree of blood loss include the number of levels fused, duration of surgery and hypothermia. Children with neuromuscular disease are at increased risk of excessive blood loss. They have more osteopenic bone and it has been suggested that the absence of dystrophin causes vascular pathophysiological changes. It is important to monitor volume status and blood loss carefully in all patients, with regular haemoglobin, platelet and coagulation estimations.

**Blood conservation**

With a large area of raw, decorticated bone and nearby rich venous plexus, scoliosis surgery can result in excessive blood loss which can exceed circulating blood volume. Coagulopathy (dilutional and consumptive) can develop and prevent haemostasis. Factors influencing the degree of blood loss include the number of levels fused, duration of surgery and hypothermia. Children with neuromuscular disease are at increased risk of excessive blood loss. They have more osteopenic bone and it has been suggested that the absence of dystrophin causes vascular pathophysiological changes. It is important to monitor volume status and blood loss carefully in all patients, with regular haemoglobin, platelet and coagulation estimations.

**Minimizing blood loss**

Exposure to allogenic blood transfusions can be reduced by techniques to minimize blood loss. Simple measures include careful positioning to avoid inferior vena cava compression, preventing hypothermia, correction of coagulopathy and good surgical technique. Compression stockings and pneumatic boots are used as thromboprophylaxis, avoiding anticoagulants.

Controlled hypotension has been shown to reduce blood loss during spinal surgery. Many methods have been described; however, a mean arterial pressure of 50–60 mm Hg can be achieved with a remifentanil infusion and volatile agent without the need for vasodilators. Hypotension and surgical manipulation may reduce spinal cord perfusion and so risk neurological injury. It is therefore important to maintain continuous neurological monitoring (see below) and an adequate haematocrit to ensure oxygen delivery.

Aprotinin has been shown to reduce blood loss during scoliosis surgery by inhibiting plasmin and preserving platelet function. Different dosing regimens are described; we use a loading dose of 4 mg kg\(^{-1}\) (28 000 KIU kg\(^{-1}\)) then infuse at 1 mg kg\(^{-1}\) h\(^{-1}\) (7500 KIU kg\(^{-1}\) h\(^{-1}\)) for the duration of the surgery. It is a bovine derived polypeptide and so can cause hypersensitivity reactions. A test dose should be given before turning the patient prone to exclude hypersensitivity and it should not be reused during staged procedures within 6 months.

**Autologous blood**

The use of autologous blood can also reduce the need for allogenic transfusions. This may be made possible by pre-donation, intraoperative acute normovolaemic haemodilution or intraoperative cell salvage.

Pre-donation of blood 2–4 weeks before surgery reduces allogenic transfusion in adolescent idiopathic scoliosis. Problems with timing of the donations with surgery and acceptance to the patient can limit its use. Smaller children and those with neuromuscular disease do not pre-donate. We use oral iron supplementation to minimize preoperative anaemia, but recombinant erythropoietin has also been used to raise preoperative haemoglobin concentrations and facilitate pre-donation.

Acute normovolaemic haemodilution can be used instead of, or alongside, autologous blood pre-donation to reduce allogenic blood transfusion. The volume of blood to be removed is calculated from the estimated blood volume and measured preoperative haemoglobin or haematocrit.

\[
\text{Volume to be removed} = \frac{\text{estimated blood volume} \times (\text{preop Hb} - \text{target Hb})}{\text{average Hb}}
\]

The blood is withdrawn from a large bore peripheral cannula or central line immediately before surgery, stored in bags with anticoagulant and labelled carefully. Normovolaemia is re-established with an i.v. infusion of colloid (ratio 1:1). The resulting blood lost during surgery has a lower haematocrit. Once haemostasis is achieved or a target haemoglobin reached, the donated blood can be transfused.

In our opinion, intraoperative cell salvage is mandatory for this type of surgery. Blood is collected, anti-coagulated, filtered, centrifuged and re-suspended in saline. Approximately 50% of blood loss can be salvaged and, in our institution, swab washing has been shown to recover a significant amount of red cells. Despite the initial outlay for equipment, disposables and training personnel, it may be cost effective with the rising price of allogenic blood.

**Neurological monitoring**

Neurological injury may occur because of direct spinal cord or nerve damage during instrumentation, distraction injury or reduced spinal cord perfusion resulting in ischaemia. Intraoperative spinal cord monitoring is used in an attempt to detect neurological injury and prevent devastating, irreversible damage.

**Continuous intraoperative neurophysiological monitoring**

The nervous system is stimulated and the response is monitored distal to the area of spinal cord at risk. This may be done using somatosensory evoked potentials (SSEPs) or motor evoked potentials (MEPs).

SSEP monitoring involves stimulating a peripheral nerve, often the posterior tibial nerve, and then detecting a response with epidural or scalp electrodes. The evoked potentials are averaged more than 2–3 min to eliminate background noise then displayed as voltage against time. Nerve injury may be indicated by decreased amplitude or increased latency. In MEP monitoring, transcranial electrical impulses stimulate the motor cortex and
the resulting signal is detected with epidural electrodes or as compound muscle action potentials (CMAPs).

Anaesthetic technique impacts upon spinal cord monitoring. Volatile agents, propofol and nitrous oxide all depress SSEPs and MEPs; however, opioids have little effect. Neuromuscular blocking agents may reduce background noise when using SSEPs, but a profound block will prevent CMAPs. Decreases in blood pressure and temperature may also depress signals. Baseline recordings are made after induction of anaesthesia and a steady state maintained to minimize drug induced changes. Any decreased amplitude, increased latency or loss of waveform must then be attributed to neurological injury. If a change in the recorded evoked potentials occurs and an injury is suspected, a ‘wake-up’ test should be performed.

**Intraoperative ‘wake-up’ test**
The ‘wake-up’ test provides a snapshot of spinal cord motor function. Surgery is halted, the volatile agent switched off and emergence allowed. The patient is asked to move their feet and, once this occurs, anaesthesia can be recommenced. Assistance is needed to prevent patient movement which may cause accidental extubation or loss of vascular cannulae. In the event of new paraplegia, all implants should be removed, hypotension and anaemia corrected, and a course of high dose methylprednisolone commenced.

**Postoperative management**

Scoliosis correction involves prolonged surgery with significant blood loss and potentially difficult postoperative pain management. Most patients are extubated immediately after surgery to enable early neurological assessment. Invasive monitoring is continued in a high dependency area. Children with neuromuscular disease or significantly reduced cardio-respiratory capacity require monitoring in an intensive care environment. Some patients may need a period of postoperative ventilation to allow volume, temperature and metabolic abnormalities to be corrected before extubation.

**Pain control**

Good postoperative analgesia is essential to allow frequent physiotherapy and early mobilization, and so reduce the risk of respiratory complications. After such extensive surgery, postoperative pain management requires a multimodal approach, combining simple analgesics, systemic opioids and regional anaesthesia.

An epidural catheter, or a paravertebral catheter during an anterior correction, can be placed intraoperatively by the surgeon. After initial neurological assessment, a loading dose of local anaesthetic is then given followed by a continuous infusion. However, because of the size of the wound and the surgical disruption of the epidural space, additional analgesia is needed.

Opioids can be administered intravenously, intrathecally or via the epidural space. We supplement an epidural infusion of L-bupivacaine 1.25 mg ml⁻¹ with a continuous i.v. infusion of morphine and regular paracetamol. Non-steroidal anti-inflammatory drugs (NSAIDs) are effective analgesics and can reduce opioid requirements. However, they may increase postoperative bleeding and interfere with bone fusion. We give NSAIDs after 24 h, or earlier in cases where pain control is difficult.

**Fluid management**

Postoperative fluid management requires careful attention. Some ongoing blood loss is likely, and paralytic ileus is possible. Decreased urine output may also result from syndrome of inappropriate antidiuretic hormone, so injudicious use of hyponatraemic fluids may result in hyponatraemia and hypomolarity. Each patient should have their volume status regularly assessed and ongoing losses should be replaced. A full blood count, coagulation studies and serum electrolytes measurement should be repeated after surgery.

**References**


Please see multiple choice questions 11–13.