Anaesthesia for paediatric eye surgery


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**INTRODUCTION**

Unlike adults, children requiring eye surgery do not tolerate sedation or local anaesthetic techniques and therefore almost always require general anaesthesia. This update will present a general review of the principles of anaesthesia for children undergoing eye surgery and a description of anaesthesia for some specific procedures.

**GENERAL PRINCIPLES OF ANAESTHESIA FOR PAEDIATRIC EYE SURGERY**

**Preoperative considerations**

Most children presenting for eye surgery are healthy, ASA I or II and may be managed as day cases. A small number have underlying conditions, often of a chromosomal or metabolic nature, which pose more specific anaesthetic challenges. Examples of these are described in appendix 1.

**Ophthalmic medications**

Many children requiring eye surgery receive eye drops. Knowledge of commonly used drugs and potential side effects is useful (See table 1). Medications may be absorbed through the pharyngeal mucosa via the nasolacrimal ducts to cause systemic effects, although this is rarely a significant problem.

**Anaesthetic considerations**

**Premedication and induction of anaesthesia**

The decision to premedicate the child and the choice of induction technique, intravenous (IV) or inhalational, should be tailored to the needs of the child and to the preferences of the anaesthetist. Children with visual impairment should be handled in a careful and sensitive manner.

**Airway management**

Airway management should be tailored to the procedure. For measurement of intraocular pressure (IOP), spontaneous respiration via a facemask should be used, as intubation will raise the intraocular pressure. For simple procedures such as examination under anaesthesia (EUA) it may be more convenient to maintain spontaneous respiration through a reinforced laryngeal mask airway (LMA), particularly where a sterile field is required.

The reinforced LMA may be used in older children for most eye procedures. It is possible to use controlled ventilation with muscle relaxants, and coughing is reduced at the end of the surgery.

Intraocular surgery requires a still eye with low intraocular pressure. The airway is best managed by intubation with paralysis and controlled ventilation.

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**Table 1. Common ophthalmologic preparations and their side effects.**

<table>
<thead>
<tr>
<th>Eye preparations</th>
<th>Indication</th>
<th>Systemic side effect</th>
</tr>
</thead>
<tbody>
<tr>
<td>Beta-blockers: Timolol maleate or Betaxolol hydrochloride</td>
<td>Glaucoma</td>
<td>Bradycardia refractory to atropine; bronchospasm in asthmatics</td>
</tr>
<tr>
<td>Carbonic anhydrase inhibitors: Acetazolamide (diamox)</td>
<td>Glaucoma</td>
<td>Metabolic acidosis, electrolyte abnormalities, allergies, including Stevens-Johnson syndrome</td>
</tr>
<tr>
<td>Antimuscarinic agents: Cyclopentolate or Atropine</td>
<td>Pupil dilatation</td>
<td>Dry mucous membranes, nausea and vomiting, tachycardia</td>
</tr>
<tr>
<td>Alpha-adrenergic sympathomimetic agents: Phenylephrine 2.5%</td>
<td>Pupil dilatation</td>
<td>Hypertension, tachycardia</td>
</tr>
<tr>
<td>NSAIDS: Diclofenac sodium, Ketorolac trometamol 0.5%</td>
<td>Pain relief</td>
<td>Potential to worsen or precipitate acute asthma</td>
</tr>
<tr>
<td>Local anaesthetic agents: Amethocaine (Tetracaine), Oxybuprocaine, Proxymetacaine</td>
<td>Pain relief or prevention</td>
<td>Local anaesthetic toxicity, particularly preterm neonates</td>
</tr>
</tbody>
</table>

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**Summary**

- Children require general anaesthesia for ophthalmic procedures/surgery, but can generally be managed as day cases.
- The oculocardiac reflex may be induced during eye surgery and risks provoking dangerous bradycardias. Prevent these by premedicating with anticholinergic agents.
- Postoperative nausea and vomiting is common after eye surgery in children.

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Similarly, eye surgery in very young children is best managed with intubation and controlled ventilation to ensure a secure airway. Access to the airway will be restricted during the surgery so it is important to secure the tracheal tube firmly. A preformed south facing RAE tube is ideal, but this may be too long in neonates; a reinforced flexible tracheal tube (ETT) may be preferable in this situation.

**Maintenance of anaesthesia**

As with induction, the choice of maintenance technique rests largely on the preferences of the anaesthetist and the availability of different agents.

The incidence of dysrhythmias is increased with halothane, particularly if there is hypercapnia and eye preparations containing atropine or adrenaline are used. Isoflurane or sevoflurane may be preferable.

Propofol has anti-emetic effects. Total intravenous anaesthesia (TIVA) with propofol reduces the risk of postoperative nausea and vomiting (PONV). Remifentanil can reduce volatile requirements.5,6

Nitrous oxide is of limited value in eye surgery as it increases PONV and diffuses into gas filled spaces. It should be avoided in vitreoretinal detachment surgery where intraocular gas bubbles of sulphur hexachloride or perfluoropropane are introduced into the eye to tamponade detached surfaces as it will cause significant rise in intraocular pressure. It should also be avoided for any patient who has undergone recent vitreoretinal detachment surgery as the bubble may last several weeks. Alternatively, if nitrous oxide was used from the start of the anaesthetic, prior to placement of the gas bubble, it will diffuse out of the bubble on completion of the anaesthetic and increase risk of re-detachment.

**Anaesthetic agents and intraocular pressure**

Normal intraocular pressure (IOP) ranges from 10 to 20mmHg. Most anaesthetic agents will decrease this. Table 3 describes the effects of commonly used anaesthetic agents on IOP.7,8,10,11,12 If serial measurements of IOP are being made, it is important to be consistent with the type of anaesthetic used on different occasions.

**Anaesthetic techniques and intraocular pressure**

Physical and physiological variables have an important effect on IOP.

<table>
<thead>
<tr>
<th>Anaesthetic agent</th>
<th>Effect on intraocular pressure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Propofol, thiopentone</td>
<td>IOP reduced by 20-30% (3-7mmHg)</td>
</tr>
<tr>
<td>Halothane, sevoflurane, isoflurane, desflurane</td>
<td>IOP reduced by 20-30% (3-7mmHg)</td>
</tr>
<tr>
<td>Opioids</td>
<td>Minimal to no effect on IOP</td>
</tr>
<tr>
<td>Ketamine</td>
<td>Dose dependent increase in IOP (minor); marked when dose exceeds 5mg.kg⁻¹</td>
</tr>
<tr>
<td>Atropine</td>
<td>No effect on IOP</td>
</tr>
<tr>
<td>Non-depolarising muscle relaxants</td>
<td>Minimal to no effect on IOP</td>
</tr>
<tr>
<td>Suxamethonium</td>
<td>Significant increase in IOP within 30 sec of administration (approx 8mmHg), effect lasts for 5-7 minutes, less if given with agents that reduce IOP</td>
</tr>
<tr>
<td>Acetazolamide, mannitol, dextran</td>
<td>Used for acute reduction of IOP perioperatively</td>
</tr>
</tbody>
</table>

Table 2. Anaesthetic agents and their effects on intraocular pressure.

Laryngoscopy, coughing, straining, crying, bucking and the process of tracheal extubation may all cause a rise in IOP. This effect may be attenuated by lidocaine 1mg.kg⁻¹ IV 3 minutes prior to intubation or extubation. Use of the LMA allows smoother induction and emergence from anaesthesia and has much less effect on IOP.13,14,35

Hypoxia and hypercapnia both increase IOP. Hypocapnia and hypothermia decrease IOP.

**The oculocardiac reflex**

The oculocardiac reflex is common during eye surgery in children, and is seen in up to 60% of children undergoing strabismus surgery. It is essential to use continuous heart rate monitoring with an ECG during eye surgery in children. The reflex takes its afferent interervations from the ophthalmic division of the trigeminal nerve, relays via the sensory nucleus in the 4th ventricle, with the efferent impulse passing through the vagus nerve.1,2

Surgical traction on the extra-ocular eye muscles or pressure on the globe causes a sinus bradycardia, and occasionally junctional rhythms, atrioventricular block, atrial ectopics or ventricular ectopics. The reflex is most commonly induced by traction on the medial rectus muscle, rather than the smaller lateral rectus muscle. The bradycardia resolves almost immediately after the stimulus has been removed and weakens with repetition of the stimulus.

Atropine 20mcg.kg⁻¹ IV or glycopyrrolate 10mcg.kg⁻¹ IV at induction will block the oculocardiac reflex. If not given at induction, it is important to have the drugs drawn up and ready to administer if bradycardia should occur.

The reflex may be attenuated by application of topical local anaesthetic agents to the eye (such as tetracaine eye drops), or by blocking the afferent limb of the reflex with a peribulbar block, although this block is not usually used in children due to the risk of globe perforation.16,17

The oculocardiac reflex is less common with sevoflurane compared to halothane, and less common with deep anaesthesia compared to light anaesthesia.18 The likelihood of significant bradycardia is doubled if hypercarbia is present, so controlled ventilation should be considered. The oculocardiac reflex is more likely to occur with rocuronium compared to atracurium.1
Children who exhibit the oculocardiac reflex are more likely to develop PONV13 and should receive an antiemetic during anaesthesia.

**Extubation and emergence from anaesthesia**

It is important to avoid coughing and bucking on the tracheal tube at the end of surgery, particularly for children who have undergone intraocular surgery. For this reason, many anaesthetists use a laryngeal mask airway for eye surgery. If a tracheal tube has been used, the child should be extubated deep, if possible. If deep extubation is contraindicated, for instance if the child has a full stomach, lidocaine 1mg.kg\(^{-1}\) IV can be given to reduce rises in IOP.

**Principles of pain relief and postoperative care**

Pain after eye surgery is usually mild to moderate and can be managed with simple analgesics such as paracetamol, NSAIDS and topical local anaesthetic agents. These may be given pre-emptively as oral preparations preoperatively or rectally/IV at induction.

Squint surgery, evisceration and vitreoretinal surgery is associated with more severe pain. Analgesia should include an opioid such as fentanyl IV, paracetamol, NSAIDS, and topical local anaesthetic if possible. Multimodal analgesia should be continued into the postoperative period, with the addition of codeine phosphate or tramadol, escalating to morphine if required. The use of opioids increases the risk of PONV and antiemetics are essential.

PONV is extremely common after paediatric eye surgery, and for strabismus surgery can be as high as 60% if no prophylaxis is given. The combination of ondansetron 0.15mg.kg\(^{-1}\) IV , and dexamethasone 0.1-0.2mg.kg\(^{-1}\) IV reduces PONV to 10% in strabismus surgery.\(^{20,21}\) It is wise to leave the IV cannula in place postoperatively where PONV may be a problem so that further antiemetics and IV fluids can be given.

Ketamine is associated with emergence phenomena and the child should be recovered in a quiet area with minimal stimulation.\(^{12}\)

Most paediatric eye procedures are treated as day cases and children may resume oral intake as soon as they are able. Occasionally PONV results in an unplanned overnight admission.

**ANAESTHESIA FOR SPECIFIC OPHTHALMIC CONDITIONS AND PROCEDURES**

**EUA and measurement of IOP**

For an examination of the eyes under anaesthesia, either an inhalational or intravenous induction technique and airway maintenance with a facemask will suffice. It may be technically easier to place an LMA for a longer EUA.

Most anaesthetic agents decrease IOP, which may potentially mask a high IOP.

Some anaesthetists advocate the use of ketamine 1–2mg.kg\(^{-1}\) IV or 5–10mg.kg\(^{-1}\) 1M for IOP measurements, as it does not drop IOP. Although it may slightly raise IOP, this may be safer than having a falsely low reading. Ketamine increases secretions so should be given with either atropine 20mcg.kg\(^{-1}\) IV or glycopyrolate 10mcg.kg\(^{-1}\) IV. Airway reflexes are maintained and instrumentation of the airway is rarely required.\(^{1,11,12}\)

Alternatively, inhalational induction may be used. The ophthalmologist should be present in the room so that the IOP can be measured as soon as the child is still. The child should not be too deeply anaesthetised, the eyes should be central and the facemask must not press on the eyes.

Regardless of the technique used, the IOP should always be measured before laryngoscopy or LMA insertion, although there is little evidence to prove that the latter significantly raises IOP. A consistent technique should be ensured if serial measurements of IOP are to be made.

**Syringing and probing of nasolacrimal ducts**

Children with blocked nasolacrimal ducts will usually present early in life with increased tearing. Most respond to probing of their nasolacrimal ducts, which is a short procedure for which an LMA will suffice.

Should simple probing fail, the surgeon might place a silicone catheter through the duct where it is secured for a few weeks. Alternatively the inferior turbinate bone may be fractured to relieve the obstruction.

Dacrocystorhinostomy is a more extensive procedure that involves exposure of the duct and creation of a new opening into the nasal cavity.\(^{1}\)

**Anaesthetic considerations**

The main problem is bleeding from the nasal mucosa:

- Topical vasoconstrictors reduce bleeding from the nasal mucosa.
- Hypotensive anaesthesia may be required to reduce bleeding, for instance, relatively deep anaesthesia with moderate head up tilt.
- The airway should be protected from blood, ideally with a throat pack, and the nasopharynx should be suctioned before extubation.
- Opioids may be required for analgesia for this procedure.

**Strabismus surgery**

Squint is a common problem that affects 3 – 5% of the population, making strabismus surgery the most commonly performed eye operation in children. It affects males and females equally.

Squints are usually idiopathic, but may also be secondary to intracerebral space occupying lesions, trauma, infection or inflammation causing muscle palsies. Most patients are healthy, but occasionally squints may be associated with a family history, prematurity, and disorders of the central nervous system such as cerebral palsy, hydrocephalus and myelomeningocele. Patients may have occult myopathies and there is a threefold increase in the incidence of maseter spasm. Anecdotal evidence of an increased association with malignant hyperpyrexia remains unproven.\(^{2}\)

Squint correction is achieved by lengthening (recession), shortening or tightening (resection) or transposition of any of the four rectus and two oblique extra-ocular muscles, or combinations of any of the above.

Surgeons may use forced ducting testing to distinguish a paretic muscle from one that has restricted motion. Botulinum toxin may be injected into the extra-ocular muscle for minor abnormalities.
This requires electromyelogram (EMG) control and muscle relaxants should be avoided. 

In older children an adjustable suture may be used that allows fine adjustments to be made 24 to 48 hours postoperatively under topical local anaesthetic once the patient is awake.

**Anaesthetic considerations**

- Induction technique, the method of airway control and choice of ventilation may be guided by the preference of the anaesthetist.
- TIVA with propofol reduces PONV. Alternatively, anaesthesia may be maintained with a volatile agent and air/oxygen.
- Consider atropine 20mcg.kg\(^{-1}\) IV or glycopyrolate 10mcg.kg\(^{-1}\) IV to block the oculocardiac reflex.
- PONV is common postoperatively, up to 50–75%. Give two anti-emetic agents such as ondansetron 0.1mg.kg\(^{-1}\) IV and dexamethasone 0.1-0.2 mg.kg\(^{-1}\) IV.\(^{20,21,22}\)
- Exutabate the child deep if possible.
- Analgesia should include topical tetracaine or oxybuprocaine, NSAIDS such as ibuprofen or diclofenac and paracetamol, unless contraindicated.
- Intraoperative opioids should be avoided if possible.
- A peribulbar block is effective for analgesic requirements and reduces PONV, possibly by blocking the ophthalmic division of the trigeminal nerve that passes to the vomiting centre in the medulla. The risk of globe perforation in children makes most practitioners cautious of this.\(^{17}\)
- A sub-Tenon block performed intraoperatively by the surgeon provides effective analgesia.

**Glaucoma**

The pressure within the eye is maintained through a balance between the production of aqueous humor, primarily by the ciliary body in the posterior chamber, and drainage via the trabecular network to the canal of Schlem in the anterior chamber.

In glaucoma the normal IOP of 10–20mmHg is elevated so that capillary blood flow to the optic nerve is reduced, which compromises the function of the optic nerve.

The causes of glaucoma are varied:
- Primary congenital glaucoma is caused by a failure of the development of the trabecular network, reducing the drainage of aqueous humour. It is bilateral in 75% of cases and has a prevalence of 1:10000 births. There is a male to female ratio of 35%: 65% and it is more common in children below the age of 3 years.
- Secondary glaucoma is usually caused by blockage of existing drainage channels by infection, inflammation or trauma. It is also seen in some rare syndromes such as Sturge-Weber Syndrome, Axenfeld Syndrome and in association with aniridia in 20% of patients with Wilm's tumour.\(^{2}\)

Treatment may be medical or surgical. Medical treatment consists of drugs used to reduce IOP. Acetazolamide 15–30mg.kg\(^{-1}\).day\(^{-1}\) PO in 3 – 4 divided doses suppresses aqueous production, but its usefulness is limited since the causes of glaucoma are usually structural and related to drainage.

**Surgical treatments may vary:**
- Goniotomy involves visualising the anterior chamber with a gonioscope and making an incision into the trabecular meshwork to allow drainage.
- Trabeculotomy involves the insertion of a fine probe into Schlem's canal to create a new drainage channel.
- Trabeculectomy involves the creation of a new drainage channel from the anterior chamber into sub-Tenon's space where the aqueous is absorbed.
- Cyclocryotherapy is the ablation of part of the ciliary body by a cryoprobe at - 60 to - 80 degrees Celsius to reduce the production of aqueous humour.\(^{2}\)

**Anaesthetic considerations**

- Avoid raising the IOP by ensuring a smooth induction and deep emergence without coughing.
- Maintain a motionless eye; consider muscle relaxants and controlled ventilation to avoid hypercapnia.
- Analgesia with paracetamol and NSAIDS is usually adequate, however when cyclocryotherapy is used opioids may be necessary.
- High incidence of PONV, give routine anti-emetics.

**Cataract extraction**

Cataracts are a major cause of childhood morbidity worldwide, predominantly in the developing countries of Africa and Asia (85% of cases).

Aetiology may be varied:\(^{2}\)
- Hereditary cataracts are autosomal dominant and present in otherwise healthy children.
- Syndromes may be associated with cataracts. Some include Lowe’s oculo-cerebro-renal syndrome (X-linked recessive), Down syndrome (trisomy 21), Edward syndrome and Cri-du-chat syndrome.
- Metabolic causes of cataract may include glucose-6-phosphate dehydrogenase deficiency, hypoglycaemia, hypocalcaemia and galactosaemia.
- Blunt or penetrating trauma over time may cause unilateral cataracts.
- Inflammation such as the uveitis associated with juvenile chronic arthritis may cause cataracts.
- Tumours, such as retinoblastoma can cause cataracts.
- Intrauterine infections, including rubella, cytomegalovirus (CMV), toxoplasma and toxocariasis.
- Radiation for leukaemia might cause cataracts.
- Chronic steroid use can result in cataracts.

Treatment involves surgical implantation of an intraocular lens. This needs to be done very early (as early as 4 weeks old) in order to allow
stimulation of the retina and visual development. The procedure takes about 30-60 minutes, but complications can be more frequent than in adults and include uveitis, glaucoma, endophthalmitis, iris damage or prolapse, retinal detachment and thickening of the posterior lens capsule.1

Anaesthetic considerations
- Aim for a motionless eye either with deep anaesthesia or muscle relaxants.
- Avoid high IOP with a smooth induction and emergence.
- Consider controlled ventilation to avoid hypercapnia.
- Give anti-emetics.

Enucleation and evisceration
Enucleation is the removal of the whole eye. This may be done for surgical treatment of a retinoblastoma, significant eye trauma or for cosmetic reasons where an eye is blind. It involves the dissection of the extra-ocular muscles off the globe. There is a similar risk here for the oculocardiac reflex as in squint surgery, although less risk of PONV. IV atropine/glycopyrrolate should be available.

Evisceration involves the removal of the contents of the globe, but retention of the sclera. This procedure is often painful and opioid analgesia will be required.1

Penetrating eye injury
A penetrating eye injury is a relatively common injury in children, primarily boys between 3 and 9 years. Surgery is required to close the defect or remove a foreign body. Up to 30% of these injuries may be associated with trauma to the head, orbit or adnexa, and the risks of anaesthesia under these circumstances need to be weighed against the benefits of early closure.23 Surgery is urgent, as anything that raises IOP (coughing, straining) may cause the globe to extrude its contents.25

This presents two conflicting anaesthetic problems. First, the child may have a full stomach so a rapid sequence intubation with suxamethonium is indicated in order to prevent aspiration. Second, there is a need to protect the globe from a rise in IOP that could result in extrusion of the structures of the anterior chamber or the vitreous humor. The transient rise in intraocular pressure produced by the use of suxamethonium could theoretically cause this.24

- One approach recommends the use of a large dose of non-depolarising muscle relaxant (NDMR) and ventilation with cricoid pressure until intubating conditions are achieved, providing the child has a normal airway.10
- Another view is that there have been no documented reports of vitreous extrusion after the use of suxamethonium, and protection of the airway is paramount;25 hence the use of suxamethonium and a traditional rapid sequence induction is indicated.

Other considerations include:
- Crying, coughing and straining should be avoided; consider light oral sedation and analgesia preoperatively.
- Direct laryngoscopy of a poorly paralysed airway can cause coughing and bucking, whichever technique is used.
- Consider blunting the intubation response prior to laryngoscopy. Administer lidocaine 1-2 mg.kg-1 IV given 3 minutes prior to rapid sequence intubation during preoxygenation.

Vitreoretinal surgery
Vitreoretinal surgery is performed for the repair of a detached retina, and although uncommon, may be necessary in children. Detachment may be primary where it is related to a defect in the retina, or secondary to an underlying illness. The surgery involves creating a chororetinal scar with cryotherapy and placing a scleral buckle towards the back of the eye, which serves to oppose the neuroretina and retinal pigment epithelium. The surgeon may place an intraocular bubble of either sulphur hexafluoride or perfluoropropane to tamponade the detached surfaces together.1,2

Anaesthetic considerations
- Avoid nitrous oxide if an intraocular gas bubble is used, both during surgery and for several weeks after. Parents should be given clear instructions in this regard for future anaesthetics.
- Controlled ventilation and muscle relaxants should be used
- This procedure is painful and analgesia including opioids should be considered.
- Anti-emetics should be used routinely
- Avoid raised IOP during extubation - usually achieved by deep extubation.

CONCLUSION
This update has reviewed the general principles of anaesthesia for paediatric eye surgery, as well as considerations for some common procedures. Key learning points include:

- Children require general anaesthesia for ophthalmic procedures and surgery, but most are healthy and can be managed as day cases.
- The oculocardiac reflex may be induced during eye surgery and risks provoking dangerous bradycardias, which can be prevented by premedicating with anticholinergic agents.
- Postoperative nausea and vomiting is common after eye surgery in children and might delay discharge if suitable prophylaxis is not given.

REFERENCES


### Appendix 1. Conditions associated with eye abnormalities requiring special precautions.

<table>
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<tr>
<th>Underlying condition</th>
<th>Eye condition</th>
<th>Special precautions</th>
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</thead>
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<tr>
<td>Neonates and premature babies</td>
<td>Congenital cataracts</td>
<td>As for neonatal surgery – warming, glucose management, monitoring for postoperative apnoea</td>
</tr>
<tr>
<td>Craniosynostosis syndromes (Crouzon, Apert and Pfeiffer syndromes)</td>
<td>Glaucoma, cataracts, squint, exophtalmos</td>
<td>Difficult to maintain airway with facemask (mid-face hypoplasia); improved by oral airway; intubation usually easy</td>
</tr>
<tr>
<td>Craniofacial syndromes (Goldenhar, Treacher Collins, Smith-Lemli-Opitz)</td>
<td>Glaucoma, cataracts, squint</td>
<td>Micrognathia/facial asymmetry – difficult intubation</td>
</tr>
<tr>
<td>Mucopolysaccharidoses (Hunter and Hurler syndromes)</td>
<td>Corneal opacities, retinitis pigmentosa</td>
<td>Difficult airway and intubation, cardiomyopathy, cervical spine instability</td>
</tr>
<tr>
<td>Down syndrome, Edward syndrome, Cri-du-Chat syndrome</td>
<td>Cataracts, strabismus</td>
<td>Difficult intubation, cervical spine instability in Down syndrome</td>
</tr>
<tr>
<td>Hallerman-Strieff syndrome</td>
<td>Congenital cataract</td>
<td>Difficult intubation</td>
</tr>
<tr>
<td>Stickler syndrome</td>
<td>Glaucoma, chorioretinoid degeneration, lens dislocation</td>
<td>Cleft palate and associated airway problems</td>
</tr>
<tr>
<td>Homocystinuria</td>
<td>Lens dislocation</td>
<td>Hypoglycaemia</td>
</tr>
<tr>
<td>Marfan syndrome</td>
<td>Lens dislocation</td>
<td>Aortic root dilatation, aortic/mitral valve regurgitation</td>
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<tr>
<td>Neuro-oculo-cutaneous disorders (Neurofibromatosis Sturge-Weber syndrome, tuberous sclerosis, Von-Hippel-Lindau syndrome)</td>
<td>Retinal vascular disorders</td>
<td>Seizures, intracranial lesions, cardiac lesions and phaeochromocytoma</td>
</tr>
</tbody>
</table>