Managing the difficult airway in the syndromic child

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Key points

- Difficult airway is 'the clinical situation in which a conventionally trained anaesthesiologist experiences difficulty with facemask ventilation of the upper airway, difficulty with tracheal intubation, or both'.
- Difficult laryngoscopy is estimated in 1.35% of paediatric procedures.
- Younger children (neonates and infants) have a higher incidence of airway management problems than older children.
- Mandibular and maxillary hypoplasia, restricted mobility in the temporomandibular joint, or fusion of cervical vertebrae contributes to potentially difficult airway management.
- Effective preparation, adequate expertise in difficult paediatric airway management, familiarity with difficult airway management algorithms, and regular refresher training are essential for success.

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A difficult airway as defined by the ASA Task Force on Management of the Difficult Airway is 'the clinical situation in which a conventionally trained anaesthesiologist experiences difficulty with facemask ventilation of the upper airway, difficulty with tracheal intubation, or both'. In a retrospective review of 11 219 paediatric procedures, the risk of difficult laryngoscopy was estimated as 1.35%. The risk was found to be higher in neonates and infants, children who are underweight, ASA physical status III and IV, or have Mallampati score III and IV.[1] However, the reliability of the Mallampati score in predicting difficult airway management in the paediatric population has been questioned and some clinicians prefer the Colorado Pediatric Airway Score (COPUR), since it uses a detailed scoring system and is therefore possibly more reliable.[2] Cooperation for airway assessment in children is not always easy and the availability of a score which accounts for a number of different aspects of the airway is more thorough. (See Table 1 for details.)

Data pertaining to the real incidence of difficult airway management in children are sparse, but they are thought to be lower than in the adult population. Certain features predicting potential difficulties with airway management are often present in a number of syndromic children seen in paediatric anaesthesia practice. Predictors of difficult intubation include the presence of dysmorphic features, limited neck extension due to fusion of cervical vertebrae as found in Klippel–Feil syndrome, limited mouth opening, and restricted mobility of temporo-mandibular joints, a large tongue (macroglossia) such as found in Beckwith–Wiedemann syndrome, limited submandibular space (retrognathia, micrognathia, mandibular hypoplasia or dysplasia) as found in Pierre Robin syndrome and Treacher Collins syndrome, and the presence of structural abnormalities in the laryngo-tracheal passage. Other predictors include soft tissue tumours, storage diseases such as mucopolysaccharidoses, and arteriovenous or lymphatic malformations involving the airway. In neonates and infants, the lateral profile may be more useful in eliciting the subtle signs of mandibular hypoplasia which are easily missed. (Refer to Table 2 for the anatomical site predominantly causing the airway problem in these syndromes.)

This review focuses on some common syndromes predominantly seen in paediatric anaesthesia practice that are associated with potentially difficult airway management and highlights the related relevant features. A broader discussion of difficult intubation in the paediatric population has been published elsewhere and is beyond the scope of this review.[3 4]

There is no shortage of techniques and devices available to the anaesthesiologist's armamentarium to successfully manage the difficult airway.[5] However, experience and familiarity with the equipment used will determine one's approach and are certainly more important than the actual device itself. This is reflected in numerous case reports citing a wide variety of strategies to successfully manage the difficult airway.

Besides the routine general preoperative assessment, the clinical evaluation of the syndromic child with a potentially difficult airway should focus on signs and symptoms of airway obstruction, including a history of apnoea episodes and daytime somnolence and also evidence of noisy breathing, stridor, snoring, and increased work of breathing. Owing to the anatomical changes, obstructive sleep apnoea syndrome is often present in these patients. The clinical examination should include a review of dentition (e.g. loose teeth, overbite), extent of mouth opening, head–neck mobility, facial anomalies (micro- or retrognathia, mid-face hypoplasia), anomalies of the palate and mandibular floor, and assessment of the thyromental distance. In addition, non-airway-related clinical problems are common in children with syndromes (e.g. congenital cardiac lesions, susceptibility to malignant hyperthermia, seizure disorders, intracranial abnormalities with

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Table 1 Details of the COPUR airway scoring[2]

Colorado Pediatric Airway Score (COPUR)	Points
C: chin	
From the side view, is the chin	
Normal size?	1
Small, moderately hypoplastic?	2
Markedly recessive?	3
Extremely hypoplastic?	4
O: opening	
Interdental distance between the front teeth	
>40 mm	1
20–40 mm	2
10–20 mm	3
<10 mm	4
P: previous intubations, OSA (obstructive sleep apnoea)	
Previous intubations without difficulty	1
No past intubations, no evidence of OSA	2
Previous difficult intubations, or symptoms of OSA	3
Difficult intubation—extreme or unsuccessful; emergency tracheotomy; unable to sleep supine	4
U: uvula	
Mouth open, tongue out, observe palate	
Tip of uvula visible	1
Uvula partially visible	2
Uvula concealed, soft palate visible	3
Soft palate not visible at all	4
R: range	
Observe line from ear to orbit, estimate range of movement, looking up and down	
>120°	1
60–120°	2
30–60°	3
<30°	4
Modifiers: add point for	
Prominent front 'buck' teeth	1
Very large tongue, macroglossia	1
Extreme obesity	1
Mucopolysaccharidoses	2

Predictions Points	Intubation difficulty	Glottic view
5–7	Easy, normal intubations	1
8–10	More difficult, laryngeal pressure may help	2
12	Difficult intubation, fibreoptic less traumatic	3
14	Difficult intubation, requires fibreoptic or other advanced methods	3
16	Dangerous airway, consider awake intubation, advanced methods, potential tracheotomy (Patients with hypercarbia awake, severe obstruction)	4
16+	Scores >16 are usually incompatible with life without an artificial airway	

raised intracranial pressure, or developmental delay), which need to be taken into consideration for the approach to the difficult airway. Previous anaesthetic history and airway management should be perused, but it should be noted that airway dynamics and dimensions may significantly change over time in some conditions. (Refer to Table 3 for the non-airway-related features that may be associated with the syndromes described below.)

The anaesthetic plan should be discussed with the child (where appropriate) and the parents/care-givers before induction of anaesthesia. Depending on the expected airway management difficulties, the possible need of establishing a surgical airway should be mentioned at this stage, particularly if surgery is considered urgent. Before commencement, a 'bail out' plan for a failure in establishing a secure, non-surgical airway ought to be in place and agreed upon by the specialities involved (refer to the Difficult Airway Society Guidelines).[3] Premedication, the choice of anaesthetic technique, checking, and setting up of special equipment will not only need in-depth consideration, but also adequate time for these issues to be fully addressed before embarking on anaesthetic induction.

Recent discussions in the literature have advocated for administration of neuromuscular blockers without prior verification of facemask ventilation in patients with normal airway. However, in syndromic patients, increased difficulty with airway management should be anticipated and hence we still recommend confirming the ability of facemask ventilation before paralysis. Once satisfactory facemask ventilation has been confirmed, neuromuscular blockade will not only improve intubating conditions, but most likely also facilitate mask ventilation.

There now follows a discussion of the airway problems associated with a selection of syndromes.

Table 2 Difficult airway in congenital syndromes based on anatomical site

Anatomical site	Related syndromes
Nasopharynx	Mucopolysaccharidoses
Oral cavity/oropharynx	Trisomy 21
	Beckwith–Wiedemann syndrome
	Mucopolysaccharidoses
Mandible/maxilla	Pierre Robin sequence
	Treacher Collins syndrome
	Goldenhar syndrome
	Apert syndrome
Pharynx/larynx	Trisomy 21
Trachea	Trisomy 21
	Mucopolysaccharidoses
Cervical spine	Trisomy 21
	Klippel–Feil syndrome
	Goldenhar syndrome
	Mucopolysaccharidoses

Trisomy 21

Trisomy 21 (T21) is the most common chromosomal abnormality occurring in 1:600–800 live births. There are multisystemic abnormalities in T21 that increase the risk of complications perioperatively.

Issues for airway management

The main airway issues in these patients are related to their short neck, relative macroglossia, microdontia, mid-facial and mandibular hypoplasia, atlanto-axial instability with vertebral ligamentous abnormalities, and a higher incidence of congenital subglottic and/or tracheal stenosis. Tonsillar and adenoidal hypertrophy often contribute to upper airway obstruction, including obstructive sleep apnoea syndrome in many T21 patients.

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Table 3 Non-airway related features of the syndromes described

Syndromes	Inheritance	Characteristics
Trisomy 21	Sporadic	Learning difficulties (99.8%) Separation of the abdominal muscles (80%) Flexible ligaments and hypotonia (80%) Congenital heart disease (45%) – Atrioventricular septal defect (40%) – Ventricular septal defect (35%) Haematological malignancies: acute lymphoblastic leukaemia and acute myelogenous leukaemia Hypothyroidism Increased risk of Hirschsprung's disease Hearing impairment (38–78%)
Beckwith–Wiedemann syndrome	Sporadic (new mutations) or inherited (autosomal dominant)	Macroglossia, omphalocoele, or umbilical hernia Gigantism at birth or post-natally Risk of hypoglycaemia (hyperplasia of pancreatic islet cells) Cardiac anomalies, Alveolar hypoventilation Skeletal anomalies Mental retardation Congenital hypothyroidism Diaphragmatic hernias Hearing loss Risk of developing malignant tumours such as – Wilm's tumour – Adrenal carcinoma – Gonadoblastoma
Pierre Robin sequence		May occur in isolation or could be associated with other syndromes
Treacher Collins syndrome	Autosomal dominant	Conductive hearing loss Drooping lateral lower eyelids and vision loss Malformed or absent ears
Goldenhar syndrome	Mostly sporadic	Scoliosis Underdeveloped or absent internal organs Limbal dermoids Hearing loss Blindness
Aperts syndrome	Autosomal dominant or sporadic	Limb anomalies
Klippel–Feil syndrome	Autosomal dominant or recessive	Abnormalities of the skeletal system – Scoliosis (60%), Sprengel's deformity (25–35%) Cardiac abnormalities (4.2–14%) – Ventricular septal defect – Patent ductus arteriosus – Mitral valve prolapse – Bicuspid aortic valve – Coarctation of aorta Genitourinary abnormalities – Absent kidney Low IQ Deafness Ocular anomalies
Mucopolysaccharidoses	Autosomal recessive and X-linked recessive (Hunter's syndrome)	Multisystem involvement based on symptoms and signs developed from deposition of glysoaminoglycans on bone, skeletal structure, connective tissues, and organs – Mental retardation – Cardiomyopathy – Hepatosplenomegaly – Obstructive sleep apnoea – Hydrocephalus

Mask ventilation might be difficult and the use of an oral and/or nasal airway is helpful. However, laryngoscopy tends to be straightforward as mouth opening is usually normal and the laryngoscope blade easily displaces the large, but soft tongue. A retrospective review of 99 patients with T21 found that one-third of the patients developed post-extubation stridor, noting an increased incidence in

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younger patients who are small for their age (less than the 5th percentile for weight) and in those who required reintubation after the initial extubation. It is estimated that congenital subglottic stenosis occurs in <1% of T21 children, but this may be an underestimate due to a lack of preoperative screening with direct laryngoscopy or bronchoscopy. Patients with T21 often have tracheal narrowing and it is recommended that a tracheal tube 0.5–1 mm smaller than the calculated size for the age of the child be used to avoid subglottic trauma.

Atlanto-axial instability in T21 is caused by the laxity of the transverse ligament and/or bony abnormalities such as a malformation of the odontoid process. The occurrence of symptoms and signs of myelopathic compromise such as motor abnormalities (change in gait and weakness of arms or legs, spasticity, hyperreflexia), a change in bowel or bladder function, significant neck or radicular pain, head tilt, or torticollis should be investigated before anaesthesia.

A child has to be 3 yr of age or older for accurate radiological imaging of the cervical spine due to ongoing mineralization process. Asymptomatic patients with plain cervical flexion and extension radiographs series showing an atlanto-dens interval of <4.5 mm and a neural canal width of more than 14 mm should be able to proceed to surgery.[6] In the presence of abnormal radiological findings, symptomatic patients, or both, full cervical spine precautions should be undertaken in the emergency situation and a referral to neurosurgery would be advisable if encountered in the elective setting. However, lateral neck radiographs may not reliably rule out atlanto-axial instability and hence are not performed routinely anymore in many centres.[7] Neck flexion-extension and rotation movements should be kept to a minimum in all these patients and proper precautions need to be taken for positioning in order to maintain the neck in a neutral position. Consider maintaining the position with the use of a soft cervical collar after induction to preserve the position of the neck, particularly for long procedures. For interventions that require extreme neck movement in symptomatic patients, an MRI investigation for spinal cord compromise would be advisable.

Beckwith–Wiedemann syndrome

This syndrome is characterized by omphalocoele, macroglossia, visceromegaly, and gigantism. It occurs in ~1:14 000 live births with an equal distribution between the sexes.

Issues for airway management

Maxillary hypoplasia and macroglossia may result in upper airway obstruction and difficult direct laryngoscopy. Obstructive sleep apnoea syndrome may be present and result in alveolar hypoventilation with subsequent pulmonary hypertension and cor pulmonale. Visceromegaly (from enlargement of the liver, spleen, and kidneys or from intra-abdominal tumours) may shift the diaphragm upwards, thereby reducing functional residual capacity and shortening the distance from front teeth to carina resulting in increased risk of endobronchial intubation.

During induction and emergence, these patients are at increased risk for upper airway obstruction. Placing the patient either prone or lateral may help relieve the obstruction if insertion of an oral and/or nasopharyngeal airway fails to resolve the problem. Intubation can be facilitated by having an assistant pull out the tongue with forceps to improve the direct laryngoscopy view. The tracheal diameter in these children tends to be larger than that of an average child of similar age and the use of cuffed tracheal tubes in these patients has been recommended to avoid exchanging the tracheal tube for size reasons. Successful intubation with the GlideScope® has been described, but it is advisable to have a fibreoptic bronchoscopy available.

Pierre Robin sequence

Pierre Robin sequence (PRS) is characterized by the presence of the clinical triad of micrognathia, glossoptosis, and a U- or V-shaped cleft palate. It occurs in ~1:8500 live births with an equal male-to-female ratio. It is considered to be a sequence, since multiple secondary abnormalities are believed to be caused by a single anomaly. Other syndromes included in PRS are Stickler syndrome, Catel–Manzke syndrome, Toriello–Carey syndrome, and Franceschetti syndrome. In 60% of the patients, PRS is associated with other syndromes.

Issues for airway management

Hypoplasia of the mandible prevents the palatal shelf from fusion between the 8th and 10th weeks of gestation resulting in retrognathia and glossoptosis, thereby causing airway obstruction of variable severity, which will dictate the optimal timing for non-surgical or surgical interventions. Some patients have respiratory and feeding problems severe enough to require surgical interventions such as tracheostomy, mandibular distraction, or glossopexy. However, in a study by Meyer and colleagues,[8] two-thirds of PRS children were successfully managed by non-surgical airway management techniques, such as prone positioning, nasopharyngeal airway insertion, temporary use of laryngeal mask airway (LMA), or tracheal intubation.

The degree of upper airway obstruction in the presence of mandibular hypoplasia has been divided into four types. In type 1, the obstruction describes true glossoptosis where the tongue touches the posterior pharynx at the level just below the soft palate. In type 2, the tongue touches at or just above the level of the soft palate, resulting in the soft palate being compressed between the tongue and the posterior pharyngeal wall. In type 3, obstruction is the result of medial compression by the lateral pharyngeal walls and in type 4, the pharynx constricts the airway in a sphincteric manner. Surgical interventions depend on the type of obstruction. Most type 1 and 2 obstructions are managed with a nasopharyngeal airway or mandibular distraction procedures, although some type 2 obstructions require relief with tracheostomy. Most type 3 and 4 patients require tracheostomy for a more definitive relief of airway obstruction.[9]

Direct laryngoscopy and tracheal intubation in neonates and infants with PRS tend to be very difficult, but generally become

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easier with age and (mandibular) growth. In the elective setting, fibreoptic intubation is the gold standard, although numerous other approaches have been used successfully including direct use of video laryngoscopes and fibreoptic intubation via LMA.

Treacher Collins syndrome

This disorder of neural crest formation involves the first and second branchial arches and is caused by a genetic mutation on chromosome 5. It occurs in ~1:10 000 live births. Sixty per cent of cases arise from new mutations. The clinical features involve the head and neck and tend to be bilateral and symmetrical.

Issues for airway management

The main craniofacial findings relevant to the anaesthetist are maxillary, mandibular, and zygomatic hypoplasia; a high arched, cleft palate, or both; small oral aperture; and abnormalities of the temporo-mandibular joint. In contrast to Pierre Robin sequence, airway management in general and intubation in particular becomes more difficult with increasing age (mainly due to decreased mandibular growth). In a retrospective review of 123 cases, the rate of failed intubations was 5% and the Cormack and Lehane grade was recorded in 97 patients, of which 53% had grade 3 and 4 views with direct laryngoscopy.[10] Of note, all patients in that series with difficult face mask ventilation became easy to ventilate after LMA insertion and no 'can't intubate, can't ventilate' scenario or emergency cricothyroidotomy or tracheostomy airway procedures were required.

Successful intubations have been described with various techniques, including blind nasal intubation, fibreoptic intubation via LMA, fibreoptic-assisted laryngoscopy, and video laryngoscopy with GlideScope® or Airtraq® laryngoscopes.

Goldenhar syndrome

This form of hemifacial microsomia, also known as oculo-auriculo-vertebral syndrome or facio-auriculo-vertebral sequence, affects the first and second branchial arch resulting in ipsi- and unilateral (90% of cases) underdevelopment of the eye, ear, nose, soft palate, lip, and mandible. The aetiology is thought to be multifactorial, but very rarely familial causes have been described.

Issues for airway management

Maintaining a seal for facemask ventilation can be awkward due to facial asymmetry. Using gauzes with self-adhesive tape to improve mask seal has been described.[11] Difficult intubation arises from a combination of asymmetrical mandibular hypoplasia, hemifacial microsomia, tracheal deviation to one side, and craniovertebral abnormalities such as the possibility of C1–2 subluxation and potentially limited neck mobility. The radiological evaluation of the cranio-facial-vertebral abnormalities using three-dimensional CT scanning has been recommended for selected patients.

The use of fibreoptic intubation via LMA, lightwand, suspension laryngoscopy, and video laryngoscopes have been used successfully in this syndrome.

Apert syndrome (acrocephalosyndactyly)

This autosomal-dominant disorder is caused by a single-gene defect on chromosome 10 and occurs in ~1:100 000 live births. It affects the first branchial arch and results in malformations of the skull, face, hands, and feet.

Issues for airway management

The premature fusion of the coronal suture and cranial base sutures leads to hypertelorism, while the reduction in antero-posterior and downward growth of the nasopharyngeal airway results in mid-face hypoplasia with narrow nasopharyngeal passages and variable degree of choanal stenosis, a high, arched palate, which is often covered with excessive soft tissue, all contributing to obstruction of air flow. These changes may progress with age making airway management, particularly facemask ventilation more difficult. Fused cervical vertebrae are found in about two-thirds of patients (mainly at C5–6 and less commonly at the C3–4 level), and depending on the limitation of neck mobility may cause problems with intubation. The aforementioned findings contribute to obstructive sleep apnoea potentially further aggravated by narrowing of the trachea secondary to fused tracheal rings (bamboo trachea), which might require the use of a smaller than anticipated tracheal tube.

The fusion of tracheal rings leads to stiffening of the trachea, thereby affecting the ability to narrow and create high-velocity airflow to clear secretions. The accumulation of secretions could produce 'monophonic' wheezing sounds.[12]

Respiratory complications in a retrospective review occurred in 10% of anaesthetic procedures. The exact reason for the increased incidence of complications in these patients is not known, but laryngo- or tracheomalacia from previous tracheostomy combined with increased tracheobronchial secretions and their limited clearance may be contributing factors. Most respiratory complications were managed with bronchodilators, increasing depth of anaesthesia, or both.[12] Not surprisingly, patients with pre-existing upper respiratory tract infections were more likely to experience respiratory complications.

Given the high risk of airway complications resulting from upper respiratory tract infection alone in these patients, it seems prudent to postpone elective procedures until symptoms related to airway secretions and infections have been optimized.

Direct laryngoscopy view can worsen after mid-face advancement surgery which is commonly carried out for cosmetic reasons. Limited mouth opening due to fibrosis or mechanical impingement of the temporalis muscle after operation can occasionally be a contributing factor.

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Klippel–Feil syndrome

This most often sporadically occurring disorder consists of the classic triad of short neck, low posterior hairline, and limited neck mobility caused by the absence or fusion of cervical vertebrae. Abnormalities at the atlanto-occipital joint and spinal canal stenosis can co-exist. Sprengel's deformity, where one shoulder sits higher on the back than the other is seen in 25–35% of patients, contributing to decreased ventilatory capacity and difficulties with surgical positioning. The syndrome occurs in ~1:40 000 live births.

Issues for airway management

Owing to the cervical (and thoracic) spine anomalies, neck motion can be severely limited, which together with micrognathia and mandibular anomalies may render airway management difficult. There is a potential risk of spinal cord injury during laryngoscopy, intubation, and positioning of the patient. Sudden rotatory head movements inducing syncope due to compromised blood supply secondary to vascular anomalies (e.g. unilateral carotid artery agenesis) have been described.[13] Careful neck movement during airway management and positioning is therefore mandatory.

The degree of airway management difficulties increases with age in this syndrome. Lightwand, LMA, and fibreoptic-assisted intubations have all been successfully used.

Mucopolysaccharidoses

This group of lysosomal storage disorders leads to the accumulation of mucopolysaccharides (glycosaminoglycans) throughout the body, including skeletal structures (dysostosis multiplex), connective tissues, and internal organs. The cause is based on the absence or malfunction of lysosomal enzymes required for glycosaminoglycan breakdown. Hurler syndrome (Type I) is the most severe and best-known form. Further details on the anaesthetic implications of mucopolysaccharidoses (MPS) are discussed in a previous *CEACCP* article.[14]

Issues for airway management

The depositions of mucopolysaccharides throughout the body lead to the changes relevant for airway management. Mask ventilation and intubation will most likely be difficult due to the coarse facial features (gargoylism) with difficult facemask fit, macroglossia, thickened nasal, oral, and pharyngeal mucosa, hypertrophy of adenoids and tonsils, hypoplastic mandible, reduced temporo-mandibular joint mobility, narrowed trachea and short, immobile, and potentially unstable neck. MPS I (Hurler syndrome), IV (Morquio syndrome), and VI (Maroteaux–Lamy syndrome) are associated with potential odontoid hypoplasia and may exhibit radiological signs of atlanto-axial subluxation, which may lead to anterior dislocation and spinal cord compression. Cardiac dysfunction is also very common and before the treatment options available today for Hurler syndrome patients, that is, enzyme replacement therapy (ERT), haemopoietic stem cell transplantation (HSCT), or both, they often died prematurely from cardiomyopathy or coronary heart disease.

Hunter and (untreated) Hurler syndrome patients are of the most challenging patients in terms of airway management. In a retrospective chart review of 214 procedures among 17 patients with MPS, difficult facemask ventilation occurred in 14% and difficult intubation in 25% of anaesthetics, respectively, while intubation failed in two cases.[15] The use of an LMA for ventilation on induction is an option, which could also be used to facilitate fibreoptic intubation.

After the introduction of HCST in 1980 and ERT in 2004, there has been evidence that these therapies have alleviated airway obstruction and pulmonary complications in Hurler syndrome. HCST was associated with a lower incidence of airway problems compared with ERT in a retrospective study.[16] The natural progression of airway obstruction in 27 patients with MPS (all subtypes) was decreased in 13 out of 14 patients who underwent successful HCST.[17]

Conclusion

Syndromes with craniofacial abnormalities can be a real challenge in terms of airway management. The key to success is effective preparation, presence of personnel with expertise in difficult paediatric airway management, regular training and familiarity with the difficult intubation equipment, teamwork, and following simple algorithms for difficult airway management. The management strategy of every case will be dictated by the history and the anatomical and functional airway as assessed on physical examination before induction of anaesthesia. The optimal management for these patients would comprise utilization of familiar equipment and modifying techniques to enable successful airway control while avoiding airway complications.

Declaration of interest

None declared.

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Please see multiple choice questions 5–8.

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