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Clinical Review

Down's syndrome and anaesthesia

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Summary

Down's syndrome is a common congenital abnormality associated with characteristic morphological features, impaired intellectual development and disorders of many organ systems with a broad spectrum of severity. Many of these, including defects in cosmetic appearance, are amenable to surgical correction. The risks of anaesthesia are increased in these children. In this article the anaesthetic implications of the syndrome are reviewed and the principles of perioperative management discussed.

Keywords: anaesthesia; congenital defects, Down's syndrome; complications

Introduction

Down's syndrome (Trisomy 21) is the commonest congenital abnormality with an incidence of 6.3 per 10 000 births (Congenital malformation statistics, notification England & Wales 1991). Children with this disorder now have a greater life expectancy, often surviving well into adulthood, and increasingly present for surgery to correct conditions both related to, and incidental to the syndrome.

In addition to the classical stigmata of Down's syndrome there is a high incidence of other congenital abnormalities many of which have important implications for the anaesthetist.

Anaesthesia and surgery carry a higher risk than in normal patients and skilful perioperative management is essential for optimal outcome. All anaesthetists can expect to come into contact with such individuals and should be aware of the potential difficulties.

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Respiratory system & airway

Children with Down's syndrome have an increased incidence of abnormalities of the respiratory system which may complicate the perioperative period.

Preoperative evidence of respiratory tract infection has been found in up to 23% and there is often a history of recurrent infection because of the relative immune deficiency associated with the syndrome (Beilin *et al.* 1981).

A degree of airway obstruction and sleep apnoea are common (Clark, Schmidt & Schuller 1980). Relevant features include a relatively large tongue, crowding of the midfacial structures, a high arched, narrow palate, micrognathia, a short broad neck and a high incidence of tonsillar and adenoidal hypertrophy.

There is an increased incidence of subglottic stenosis in children with Down's syndrome (Steward 1970). Frequently a smaller tracheal tube than would have been predicted by age is required, although these children tend to be of smaller stature than normal children (Kobel, Creighton & Steward 1982). If sedation or a technique of anaesthesia involving

Table 1
Cardiac anomalies in Down's syndrome

	%
Endocardial cushion defects	40
VSD	27
PDA	12
Fallot's tetralogy	8
Others	13

From Kobel M., Creighton R.E., Steward D.J. (1982).

spontaneous ventilation is used, the patient should be carefully monitored as hypoventilation frequently occurs (Rautiainen & Meretoja 1994). Recovery may be complicated by airway difficulties and atelectasis of the lung and postoperative respiratory tract complications occur much more frequently than in normal individuals (Beilin *et al.* 1981; Kobel *et al.* 1982). Patients who have had abdominal or thoracic surgery are at particular risk of postoperative respiratory tract infections and benefit from regular physiotherapy and close monitoring. The incidence of postextubation stridor following prolonged ventilation is 30%–40% as opposed to 2% in other children (Sherry 1983), which is why some centres administer a short course of steroids prior to extubation of Down's syndrome patients.

The cardiovascular system

An association between cardiac anomalies and Down's syndrome was first described by Garrod at the end of the 19th century. The overall incidence is now estimated at approximately 40% (Rowe & Uchida 1961; Smith 1982).

The majority of congenital cardiac defects seen in Down's syndrome involve a left to right shunt. This leads to increased pulmonary blood flow and

Table 2
Clinical features of Down's syndrome

	Features	%
General	low birthweight	
	short stature	
Cardiovascular	congenital heart disease	40
	increased susceptibility to pulmonary hypertension	
	atropine sensitivity reported	
Respiratory	high arched narrow palate	70
	macroglossia	50
	micrognathia	
	increased susceptibility to respiratory infections	
	subglottic stenosis	
	postextubation stridor	30–40
	upper airway obstruction / sleep apnoea	
Gastrointestinal	dental abnormalities	65
	duodenal obstruction	8
	gastro-oesophageal reflux	
	Hirschsprung's	
Nervous system	mental retardation	99
	epilepsy	5–10
	strabismus	14–33
Musculoskeletal	hypotonia	21–80
	hyperextensibility or flexibility	47–77
	dysplastic pelvis	67
	atlanto-axial subluxation	12–32
Immune system	immunosuppression	1 in 95
	leukaemia (ALL / AML)	20 × risk
Haematology	neonatal polycythaemia	
Endocrine	low circulating catecholamine levels	
	hypothyroidism	

therefore predisposes to pulmonary hypertension. The increased incidence of pulmonary vascular disease and earlier development of pulmonary hypertension in Down's syndrome in comparison with other children with similar cardiac anomalies is well documented (Greenwood & Nadas 1976). The exact mechanism is uncertain but may relate to chronic hypoxia secondary to recurrent respiratory tract infections, upper airway obstruction and obstructive sleep apnoea (Wilson, Hutchins & Neill 1975; Rosengart & Isabel-Jones 1976). Pulmonary hypertension may develop even in the absence of congenital heart disease in Down's syndrome and Eisenmenger's haemodynamics may considerably complicate the perioperative management of an older child (Chi & Krovetz 1975).

Children with Down's syndrome undergoing cardiac surgery have a higher mortality and morbidity than age matched controls having similar surgery. The duration of postoperative ventilation, intensive care stay and total period spent in hospital all tend to be longer than for other children (Morray, MacGullivray & Duker 1986).

Immunity and haematology

Individuals with Down's syndrome have a relative immune deficiency (Albin 1978) and there is an increased susceptibility to all infections in particular those of the respiratory tract. Strict asepsis is advisable for invasive procedures and venous and arterial cannulas, urinary catheters etc. should be removed as early as possible postoperatively to minimize the possibility of serious infection. There is an increased incidence of positive hepatitis B surface antigen in this group. Polycythaemia is more common in the neonatal period (Lappalainen & Louvalainen 1972) and leukaemia (acute lymphoblastic and myeloblastic) has a frequency twenty times that seen in the general population.

Endocrine abnormalities

Down's syndrome has been linked with organ specific autoimmune dysfunction. In particular there is an increase in the incidence of congenital hypothyroidism and up to 40% of adults with Down's syndrome have evidence of hypothyroidism (Dinani & Carpenter 1990; Pozzan *et al.* 1990). Physical signs may not be helpful and biochemical screening of thyroid function is generally recommended. Thyroid antibodies are found in up to one-third of patients. Hypothyroidism should be corrected with thyroxine and although therapy appears to have little or no impact on intellectual performance (Baxter *et al.* 1975; Tirosh *et al.* 1987) it clearly has important anaesthetic implications. Relatively low blood catecholamine levels have also been reported (Keele *et al.* 1969).

Gastrointestinal system

These are the second most common group of congenital abnormalities after cardiac defects and a frequent reason for surgery early in life. The association of duodenal atresia and Down's syndrome is well known and was first described in 1949 (Lanman 1949; Grove & Rasmussen 1950; Bodian *et al.* 1952). Hirschsprung's disease is also more frequent and gastro-oesophageal reflux is also found more often in this group (Weesner & Rosenthal 1983).

Central nervous system

A degree of mental retardation is universal in Down's syndrome, however these children are generally regarded as gregarious and friendly. Nonetheless, there is a higher incidence of postoperative agitation which may occasionally warrant sedation and children may be very difficult to manage in the recovery period.

Muscle tone is abnormal in the majority of individuals with Down's syndrome and hypotonia has been reported in up to 75% which may affect the ability to maintain an adequate airway following anaesthesia.

Strabismus is more common than in normal children (14–33%) and infants commonly present for squint surgery.

Epilepsy is present in up to 10% of individuals.

Atropine in Down's syndrome

Atropine sensitivity has been reported in this group of patients. Both an exaggerated mydriatic response to ocular atropine (including death following conjunctival instillation) and an increased response of the heart rate to parenteral atropine have been documented (Berg, Brandon & Kirman 1959; Priest

1960; Harris & Goodman 1968). Although an increased mydriatic response is confirmed subsequent studies have not demonstrated cardiac sensitivity (Mir & Cumming 1971; Wark, Overton & Marian 1983). Atropine is an antisialogue and may be useful in reducing intra and postoperative airway complications, which are prevalent in this group. Vagal blockade is theoretically desirable since they have diminished sympathetic nervous system activity and a lower level of circulating catecholamines (Keele *et al.* 1969).

Atlantoaxial instability

An association between atlantoaxial instability and Down's syndrome is well documented (Spitzer, Rabinowitch & Wybar 1960; Tishler & Martel 1965). The reported incidence varies between 12% and 32% of individuals with the syndrome depending on the definition of instability and the age of the cohort (it is more common in children) (Semine *et al.* 1978; Pueschel, Scola & Perry 1981; Pueschel & Scola 1987; Elliott, Morton & Whitelaw 1988). The instability is due to laxity of the transverse atlantal ligament, which is responsible for holding the odontoid peg close to the anterior arch of the atlas. There is also a high incidence of bony abnormality of the atlas and axis (6%) which may increase the potential for instability. The normal atlantoaxial interval, the distance between the odontoid peg and the anterior arch of the atlas on lateral cervical spine x-ray, is less than 4.5 mm. Subluxation narrows the diameter of the cervical canal and may cause spinal cord compression. Most patients with an atlantoaxial distance of less than 6 mm are asymptomatic but an interval of greater than 7 mm is almost always associated with neurological manifestations (Pueschel *et al.* 1984). These include: abnormal gait, increased clumsiness, walking fatigue or a new preference for sitting games. Physical signs such as hyperreflexia, clonus, quadriparesis, extensor plantars, neurogenic bladder, hemiparesis, ataxia and sensory loss may also be present.

General concern about atlantoaxial instability in Down's syndrome increased following recommendations by the Special Olympics Committee (Special Olympics Bulletin 1983) and the Committee on Sports Medicine of the American Academy of Pediatrics (1984), that *all children* with Down's syndrome should have cervical spine x-rays prior to participation in any sporting event, and should be excluded from certain sports if there is an increased atlantoaxial interval or bony abnormality of the 1st or 2nd cervical vertebrae. These recommendations were endorsed by the Royal Society for Mentally Handicapped Children and Adults (MENCAP) in the United Kingdom but the Department of Health and Social Security and British Orthopaedic Association however, have suggested that routine screening is only justified for those individuals engaging in high risk sports (Department of Health and Social Security 1986; Royal Society for Mentally Handicapped Children and Adults 1986).

Symptomatic atlantoaxial instability in Down's syndrome was reviewed by Davidson after these recommendations (Davidson 1988). He concluded that x-ray criteria of atlantoaxial instability are *not predictive* of a tendency to dislocation. He noted that in individuals with Down's syndrome who suffered dislocation the injury was usually preceded by neurological symptoms or signs (28/31 cases). A recent prospective study of 400 children and young adults with Down's syndrome assessed the risk of spinal cord compression in individuals with x-ray evidence of atlantoaxial instability participating in potentially risk sporting activities (Cremers *et al.* 1993). After a year, there was no difference between the motor function of the group continuing high risk activities and those who had been restricted. Some had new neurological signs but these were unrelated to atlantoaxial interval at the start of the study. These findings support Davidson's suggestion that x-ray evidence of atlantoaxial instability does not correlate with risk of dislocation and cord injury.

Atlantoaxial instability is of special concern to the anaesthetist since predisposition to subluxation may be increased by the loss of muscle tone which occurs during anaesthesia. Furthermore manipulation of the head during laryngoscopy, internal jugular vein cannulation, transfer to and from the operating table and positioning for some types of surgery may carry particular risk and should all be cautiously undertaken.

There have been several case reports of spinal cord injury due to atlantoaxial subluxation in patients with Down's syndrome undergoing general anaesthesia (Moore, McNicholas & Warran 1987; Williams *et al.* 1987; Msall *et al.* 1990; Litman & Perkins 1994).

In three of these there were preoperative features which might have alerted clinicians to the possibility of atlantoaxial instability and cord compression but cervical spine x-rays had not been requested.

The question of which children should have cervical spine x-rays prior to anaesthesia remains controversial. Some authors (Williams *et al.* 1987; Elliott, Morton & Whitelaw 1988; Powell, Woodcock & Elliscombe 1990) suggest that they are mandatory in all cases, whilst others (Kobel, Creighton & Steward 1982; Moore, McNicholas & Warran 1987) feel that unless there are abnormal neurological findings in the history or examination, avoidance of forced flexion, extension or rotation of the neck under anaesthesia should be sufficient. The most recent evidence suggests that cervical spine x-rays may be unreliable in the assessment of the risk of atlantoaxial distribution. If there are neurological symptoms or signs, preoperative x-rays and orthopaedic or neurosurgical referral *are* mandatory but in their absence routine screening is not appropriate. This view is endorsed by the authors of a paper published in this issue of *Paediatric Anaesthesia* which is supported by their review of the practice of North American members of the Society of Pediatric Anesthesia (Litman, Zerngost & Perkins 1995). The authors go on to recommend that protocols be developed to improve the management of patients with atlantoaxial instability.

In conclusion, the safe anaesthetic management of children with Down's syndrome depends on an awareness of the multisystemic nature of this condition, and the potential for serious complications in the perioperative period. Neurological status in particular, should be carefully monitored because of the ever present risk of atlantoaxial instability.

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