EMERGENCY CASEBOOK

Stridor and Down's syndrome

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A 36 year old woman with Down's syndrome and hypothyroidism presented to the emergency department with breathlessness. She had suffered with coryzal symptoms for a week, before becoming increasingly dyspnoeic the day before presentation. On arrival she was unable to speak and stridulous. She was neither drooling nor toxic. Her temperature was 37.4°C. However oxygen saturation was reduced and arterial blood gas measurement showed a type 2 respiratory failure with acidosis. An urgent anaesthetic opinion was sought, and the patient transferred to the resuscitation room. Fiberoptic laryngoscopy confirmed the suspicion of a supraglottitis. After inhalation induction using sevoflurane, intubation was achieved with difficulty. Using bougie guidance, a size 7.0 orotracheal tube was found to be too large, hence the patient was intubated with a size 6.0 orotracheal tube. Cefuroxime and dexamethasone were given, and the patient was admitted to the intensive care unit. The supraglottic inflammation was monitored by regular laryngoscopy. After seven days without significant reduction in supraglottic inflammation, formal tracheostomy was performed. Blood cultures were negative.

DISCUSSION

Infection and inflammation of the supraglottic structures may involve the epiglottis only or extend to the aryepiglottic folds and arytenoid soft tissues, causing epiglottitis and supraglottitis respectively. Although widely reported in paediatric practice before routine *H influenzae* B immunisation, supraglottitis may be under-diagnosed in adults.¹ When such patients present with signs or symptoms of respiratory distress, there is risk of life threatening complete airway obstruction. Early interventional support of the airway must be considered.² A wide variety of infections have been associated with the problem including Gram positive cocci, meningococcus, pasteurella, serratia, and herpes simplex virus. Hence a broad spectrum antibiotic is needed in the first instance. In respiratory distress, temporising measures such as humidified oxygen and helium-oxygen mixtures are suggested to be useful before formal intubation is achieved. Nebulised epinephrine and corticosteroids are often used although neither is definitively proved to be of benefit.³ Emergency cricothyroidotomy may be necessary. UK consensus among emergency anaesthetists suggests inhalation induction using sevoflurane as the best way of achieving intubation.⁴

In this case Down's syndrome provided a number of additional challenges. People with Down's syndrome are prone to respiratory infections. In addition airway problems such as obstructive sleep apnoea are common, a product of craniofacial differences in anatomy. These differences, such as macroglossia and a narrow nasopharynx, may also make intubation difficult. Studies of anaesthesia for elective surgery in Down's syndrome have reported problems with atlanto-axial instability and tracheal stenosis. It is suggested that in the patient with Down's syndrome an endotracheal tube smaller than predicted should be routinely used.⁵

LEARNING POINTS

This case highlights the importance of considering supraglottitis in the adult patient with stridor or painful dysphagia. Early airway intervention may be necessary. Furthermore, the anatomical differences associated with Down's syndrome may have an important effect on disease progression and patient outcome. Emergency staff should be aware that patients with Down's syndrome may require expert intubation with a smaller than expected endotracheal tube even in the absence of supraglottic swelling.

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