Perioperative management of patients with Cockayne syndrome – recognition of accelerated aging with growth arrest

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Sir—We report the anesthesia management of a 15-year-old girl with Cockayne syndrome (CS) who presented for excision of posterior capsular membrane of the right eye. She had several characteristic diagnostic features including growth failure, weighing only 15 kg, neurodevelopmental delay, cataracts, and sensorineural deafness (1). Her preoperative blood tests and ECG were within normal limits but the glomerular filtration rate was found to be 90 $ml \cdot min^{-1}$ per 1.73 m^2. An older male sibling with CS had died at 16 years of age from progressive renal failure, diabetes mellitus, and respiratory complications.

The patient was premedicated with oral glycopyrrolate (25 $\mu g \cdot kg^{-1}$). Anesthesia was induced with sevoflurane in nitrous oxide and oxygen following which intravenous access was established. Mask ventilation was not difficult. Direct laryngoscopy with a Robertshaw blade (size 0) and application of cricoid pressure enabled a grade 2 view of the glottis. A 4.0-mm oral RAE tracheal tube was then successfully placed with a leak at 12 cmH_2O. Subsequent perioperative course was uneventful.

Cockayne syndrome is a rare autosomal recessive disorder, believed to be caused by a defect in the transcription-coupled repair of DNA (2). While there is significant variability in the clinical presentation one consistent feature is postnatal growth failure described as 'cachetic dwarfism'(1,2). Belonging to a group of progeroid syndromes, CS is characterized by variable acceleration of normal aging. Cells cultured from CS patients exhibit increased sensitivity to agents known to damage DNA, such as UV and ionizing radiation. The accumulation of DNA damage from diminished DNA repair capacity leads to transcriptional failure that may play a role in the process of aging (3). Cardiovascular diseases, such as hypertension and renal impairment are also reported (2). Signs and symptoms noticed first in infancy are progressive with the severe cases rarely surviving into adulthood. The diagnosis is established by clinical criteria, as in our patient (1). Genetic testing for the defective CSA (CS type I) and CSB (CS type II) genes or biochemical tests to quantify deficient recovery of RNA synthesis after exposure to UV-C radiation are also performed (2,4).

Few cases of anesthesia management of CS have been reported (5–9). The significant implications that have been reported include possible difficult airway with subglottic narrowing and coronary artery disease. Our patient had been anesthetized five times in our institution since she was 11 years old for various ophthalmic procedures. She never had any symptoms suggestive of gastroesophageal reflux, another feature of this syndrome, and anesthesia had previously been induced successfully with sevoflurane. On all occasions mask ventilation was not noted to be difficult and the documented difficulty in managing her airway were limited neck extension with kyphoscoliosis, anterior larynx, and the need for cricoid pressure to achieve a grade 2 view of the larynx. She had always required a 4.0-mm tracheal tube, which was associated with a large leak. Insertion of a 4.5-mm tube had either been unsuccessful or was considered too tight fitting. However, a cuffed 4.0 mm tube was used successfully. It is interesting that others have reported the use of size 3.5–4.0 mm tracheal tubes in adolescents with CS. Another significant feature was that from the age of 11–15 years she weighed between 12 and 13 kg. It is likely that because of growth failure and accelerated aging, our patient had reached the 'adult' plateau in development. Her 'adult' airway was also better served by a cuffed tracheal tube albeit one with a smaller diameter commensurate with her somatic size.

In conclusion, the primary concerns during the perioperative management of CS patients are characteristic growth arrest, i.e., failure to grow beyond an infant's size coupled with accelerated aging. Considerations for airway management should include the use of weight-appropriate rather than age-appropriate airway equipment (9). Depending on the severity of the syndrome they may reach the 'adult' plateau in their development in early childhood when it is not uncommon to encounter 'adult' diseases, such as myocardial ischemia (7), renal impairment, and diabetes mellitus. Recognition of advanced physiological age vis-à-vis their somatic appearance is essential for successful management.

Sreekrishna Raghavendran md, dnb, frca,
Karen A. Brown md,
Natalie Buu md, frcpc,
Department of Pediatric Anesthesia,
Room C-1112, Montreal Children's Hospital,
2300 Rue Tupper, Montreal, Canada
(email: sreeki71@hotmail.com)

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