

Anaesthetic considerations in a prematurely born infant with congenital hypothyroidism presenting for cataract surgery

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Abstract

Anaesthetic management of the premature infant is a challenge. This is owing to the immaturity of his or her organ systems and the possible presence of sequelae of premature birth, such as bronchopulmonary dysplasia, apnoea, patent ductus arteriosus and intraventricular haemorrhage. The premature infant is at risk of developing postoperative apnoea, hypothermia and hypoglycaemia until his or her postgestational age is more than 60 weeks. Conditions such as congenital bilateral cataract and congenital hypothyroidism may be associated with other birth defects and syndromes. Recommendations for early cataract surgery may necessitate anaesthetising premature infants with postconceptional ages less than 60 weeks, or those who have had recently diagnosed medical conditions, such as hypothyroidism. We describe the anaesthetic considerations in a, so far, unreported scenario of a premature infant with bilateral congenital cataract and congenital hypothyroidism who presented for cataract surgery.

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Introduction

Improved perinatal care has increased the survival rate of preterm babies. These survivors, with their unique problems, present for different surgeries.¹ To date, there has been no report of a premature infant with congenital hypothyroidism (CH) presenting for bilateral cataract surgery. We present the anaesthetic considerations for such a patient.

Case history

A 13-week-old male infant, posted for cataract surgery, was the first child of consanguineous parents. The mother, who had not attended any antenatal check-ups, delivered at 31 weeks in a primary care centre. She could not recall the birthweight, but felt that there had been no excessive weight gain since birth. There was no history of previous apnoeic episodes, seizures, jaundice or respiratory infection, previous hospital admission or feeding difficulties. During an immunisation visit at the postnatal age of 10 weeks, bilateral opacity of the eyes was noticed and the infant was referred to our hospital.

Since this infant had not been screened at birth, serum thyroid-stimulating hormone (TSH) was checked. A TSH

value of 750 µIU/ml prompted further workup. The total T4 and free T4 levels were not detectable. An ultrasound of the neck revealed that the thyroid gland was absent. The infant had been started on tablet levothyroxine two weeks before the preanaesthetic check-up (25 µg/day, three days a week, and 12.5 µg/day, two days a week), crushed and applied on the tongue in the morning, half an hour before starting feeds.

The infant weighed 1 700 g, appeared to be dull and had a large tongue. He was afebrile, with a heart rate (HR) of 135/minute and a respiratory rate of 35/minute. Systemic examination and reflexes were normal. Haemoglobin was 10 g/dl. Echocardiography was normal. Anaesthesia clearance was given for cataract surgery as American Society of Anesthesiologists class III. A paediatric intensive care unit (PICU) bed was arranged. The fasting orders required breastfeeds and water to be withheld for four and two hours, respectively, before the scheduled time of surgery. The morning dose of levothyroxine was given two hours preoperatively. No premedication was given.

Monitors included pulse oximetry [oxygen saturation (SpO₂)], electrocardiography, noninvasive blood pressure (BP), end-

tidal carbon dioxide and temperature. Preinduction HR was 148/minute and SpO₂ was 98%. A convective warming device and an intravenous fluid warmer were used.

Anaesthesia was induced with 100% oxygen and 4% sevoflurane, 24 G peripheral intravenous access obtained, fentanyl 4 µg given, sevoflurane increased to 6%, and the trachea intubated. Anaesthesia was maintained with 2.5% sevoflurane in 60% oxygen. The lungs were ventilated with a Mapleson F circuit. The cataract surgery on the right eye and intraocular lens implant lasted 30 minutes. The intraoperative HR was 155-170/minute, the SPO₂ was 95-99%, systolic BP was 55-75 mmHg, diastolic BP was 30-40 mmHg and the patient's temperature was 36.5-37°C.

At the end of surgery, the infant was transferred to the PICU for elective ventilation. Spontaneous respiration was resumed within two hours. Later, when he was awake with good motor activity, the trachea was extubated. Subsequently, he was sent to a step-down care facility for 12 hours and discharged after 48 hours. A similar procedure was followed for surgery on the left eye after two weeks.

Discussion

Anaesthesia is challenging in a preterm infant. Organ system maturity depends on the postconceptional age. There may be sequelae of premature birth, such as those due to birth asphyxia, respiratory distress syndrome, pulmonary hypertension, bronchopulmonary dysplasia, necrotising enterocolitis, intraventricular haemorrhage, patent ductus arteriosus, gastro-oesophageal reflux and apnoeic spells. In addition, the premature infant is at risk of developing postoperative apnoea, hypothermia and hypoglycaemia.¹

Postoperative apnoea is more likely if the infant is known to have had previous apnoeic episodes, chronic lung disease, neurological problems or a haemoglobin less than 10 g/dl, and when the postconceptional age is less than 60 weeks. General anaesthesia, using neuromuscular blocking agents and opioids, has a greater incidence of postoperative apnoea than regional anaesthesia.² The use of desflurane or sevoflurane does not decrease the incidence.³ Ideally, elective surgery should be deferred until the postconceptional age is more than 60 weeks. If surgery is required earlier, postoperative monitoring is needed for at least 12 hours.²

CH has an incidence of 1:1 000 to 1:3 500 in newborn babies, showing geographical variation. It may be permanent or transient. Developmental abnormalities of the thyroid are the most common cause (80-85%) of permanent primary CH. Thyroid agenesis and hypoplasia account for approximately 30% of this group.⁴ Thyroid agenesis is twice as common in females, than in males.⁵ Thyroid dysmorphogenesis accounts for 10-15% of permanent

primary CH. Permanent secondary (TSH-deficient) and tertiary CH (TSH-releasing, hormone-deficient) are very rare, with incidences of 1:60 000 and 1:140 000 respectively.^{4,5} Peripheral hypothyroidism is due to the defects of thyroid hormone transport, metabolism or action. CH may also be a consequence of maternal iodine deficiency or excess (drugs, contrast agents, antiseptic solutions or eating seaweed). CH may be transient in babies who are born to mothers on antithyroid drugs, or those having TSH receptor-stimulating and/or blocking bodies. Graves' disease in the mother may cause thyroidal dysfunction (hypo- or hyperthyroid) in her offspring. Premature, low-birthweight or sick babies may have transient CH.⁴

Some of the manifestations that may be present include prolonged jaundice, feeding difficulties, lethargy, umbilical hernia, macroglossia, constipation, a cold or mottled skin, abnormal crying, oedema, a puffy face, delayed reflexes and hypotonia. Approximately 30% of cases may have increased birthweight. However, most newborn babies are asymptomatic (there is the presence of maternal thyroid hormones or some functioning thyroid tissue), hence screening by measurement of serum TSH levels is the recommended method.^{4,5} Serum T4 is expected to normalise within 1-2 weeks once treatment with levothyroxine has commenced. Serum TSH normalises after four weeks.⁶ The following conditions could be considered in the differential diagnosis of symptomatic babies: growth hormone deficiency, constitutional growth delay, malabsorption syndromes, malnutrition and other causes of constipation.⁵

Infants with CH have a higher frequency of extrathyroidal congenital malformations than that of the general population. The most common are cardiac, neural and ocular anomalies. CH may also be associated with Down's syndrome. The syndromes in which hypothyroidism features include Pendred syndrome (deafness, goitre and hypothyroidism), Bamforth-Lazarus syndrome (hypothyroidism, cleft palate and spiky hair), ectodermal dysplasia (hypohidrotic, hypothyroidism and ciliary dyskinesia), Kocher-Deber-Semilange syndrome (muscular pseudohypertrophy and hypothyroidism), benign chorea-hypothyroidism, choreoathetosis (hypothyroidism and neonatal respiratory distress), and obesity-colitis (hypothyroidism, cardiac hypertrophy and developmental delay).⁶ Hirsch-sprung's disease, Beckwith-Wiedemann syndrome, Pierre Robin sequence, Albright's osteodystrophy, VATER association and frontonasal dysplasia were found to be associated with CH in a Mexican study.⁷

Patients with hypothyroidism may have depressed myocardial function, decreased spontaneous ventilation, abnormal baroreceptor function, anaemia, hypoglycaemia, reduced plasma volume, hyponatraemia, hypothermia and impaired hepatic drug metabolism. They are at

risk of developing myxoedema coma. It is advisable that a euthyroid state is achieved preoperatively. With hypothyroidism, complications during anaesthesia, such as circulatory depression, a faster inhalation induction, potentiation of anaesthetic agents and neuromuscular blockade and hypothermia, should be expected.⁸

Cataracts account for 12% of India's blind children.⁹ Congenital cataracts cause severe visual impairment because of early pattern deprivation during the sensitive period of visual development.¹⁰ Surgery that is performed before the baby reaches 10 weeks of age results in a greater chance of good vision.¹¹ Hence, there is a sense of urgency when operating on congenital cataracts.

In a study conducted in south India, 110 out of 514 children (21.4%) had bilateral congenital cataracts. Twenty-five per cent were hereditary, 15% were due to congenital rubella syndrome, and in 51% the cause could not be determined.¹² Certain syndromes may be associated with cataracts, such as Down's syndrome, aniridia, juvenile chronic arthritis with uveitis, diabetes, galactosaemia, craniosynostosis, Hallerman-Streiff syndrome, Smith-Lemli-Opitz syndrome, X-linked dominant chondrodysplasia punctata, achondroplasia, Marshall syndrome, mosaic trisomy 8, partial duplication of chromosome 1, Lowe syndrome, unspecified collagen disorder, Goldenhar syndrome, Treacher-Collins syndrome and hereditary hyperferritinaemia cataract syndrome.¹³

Anaesthetic evaluation of our patient included a meticulous search for associated conditions, the involvement of other organ systems and airway abnormalities. There were no associated syndromes or other birth defects. Since our patient's postconceptional age was 44 weeks, his haemoglobin was 10 g/dl, and surgery was to be performed under general anaesthesia, postoperative apnoea was anticipated.² Furthermore, he had been on thyroid hormone replacement therapy for a period of two weeks, which has been shown in studies to be adequate in correcting T4 levels.⁶ However, in order to maximise the benefit of early cataract surgery, the infant was accepted to undergo general anaesthesia with increased risk.

The risk of prolonged neuromuscular blockade in hypothyroidism and postoperative apnoeic spells because

of prematurity prompted us to avoid neuromuscular blocking agents. Tracheal intubation was accomplished with sevoflurane.¹⁴ The infant was extubated in the PICU once he was wide awake and monitored postoperatively for more than 12 hours in a step-down facility.²

Conclusion

Recommendations for early cataract surgery may necessitate anaesthetising premature infants with postconceptional ages less than 60 weeks, or with recently diagnosed medical conditions, such as hypothyroidism. A diagnosis of congenital cataract and/or congenital hypothyroidism should prompt a search for associated syndromes and birth defects. Careful preoperative assessment and planning are a prerequisite for a safe anaesthetic outcome.

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