Clinical Reports

Dental anaesthesia for a child with complete amelia

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A four year-old boy born without limbs (amelia) presented for dental restorations under general anaesthesia as an outpatient. Following intramuscular atropine administration anaesthesia was induced using halothane, oxygen and nitrous oxide inhaled by mask. Next, intravenous access was secured by external jugular vein catheterization. Because of his small mouth, hypognathic mandible, arched palate and anterior-superiorly located larynx, oral intubation under deep anaesthesia during spontaneous ventilation was difficult. The epiglottis was noted to be inverted on subsequent laryngoscopic inspection after intubation but was reduced mechanically to anatomic position. Despite being unable to accurately monitor the blood pressure the intraoperative period was uneventful. Postoperatively the patient was extubated and was able to return home the same day.

Complete absence of limbs is referred to as amelia or ectromelia. It represents the extreme end of a wide spectrum of congenital musculoskeletal deficiencies and deformities. The anaesthetist involved in surgery for such a patient is confronted by both the technical problems posed by the planned operation and by the difficulties of inducing and adequately monitoring the patient who may also have anomalies of other organ systems.

Key words
ANAESTHESIA: paediatric; CONGENITAL ANOMALIES: amelia.

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Case report

The patient is a four-year-old boy who presented to the Daycare Surgery Department of The Hospital for Sick Children for restoration of carious teeth under general anaesthesia. No prior notification of the patient had been given to the Department of Anaesthesia. The child had complete amelia and had resided in a children's institution in Hong Kong until his adoption by Canadian parents in 1984.

Records dating to his birth were unavailable. It was revealed from his Out-Patient Chart that his natural mother had two abortions attempted in the first and early second trimesters of her pregnancy which yielded some products of conception. A limbless male neonate was delivered by Caesarean section.

The child also had a micropenis with its undersurface fused to an incompletely formed scrotum empty of testicles. He had been clinically and cystoscopically investigated under local anaesthesia by a urologist at The Hospital for Sick Children and had been started on human chorionic gonadotrophin (HCG) to promote testicular descent.

He had undergone previous dental work under general anaesthesia in Hong Kong but no record of this was available. The patient's adoptive mother noted that his usual oral temperature was 38° C. No other anomalies had been detected.

Physical examination revealed a 7.55 kg boy whose rectal temperature was 38° C, respiratory rate was 30 per minute and heart rate was 130 per minute and regular. He was absent of even rudimentary appendages and had a smoothly rounded torso. Strong carotid and temporal pulses as well as weak ilio-inguinal pulses were palpated bilaterally. He had a small mouth with a high arched palate as well as a hypognathic mandible and an anterior-superiorly placed larynx. This patient was not sent to our

Anaesthesia Clinic and presented to anaesthesia one-half hour before the scheduled time of surgery. A urine sample was not available and because he was very upset, it was decided that a blood sample for haemoglobin determination and an expressed urine sample for protein and sugar determination would be taken after the induction of anaesthesia.

No premedication was given. After giving 0.15 mg of intramuscular atropine and attaching an electrocardiogram and precordial stethoscope, anaesthesia was induced with nitrous oxide (4 L), oxygen (4 L) and halothane by mask. A 20-gauge venous cannula was then inserted into his right external jugular vein and an intravenous solution was started (Figure). Several unsuccessful attempts were made bilaterally to insert an ilio-inguinal arterial line percutaneously to measure blood pressure and to allow blood sampling. It was decided not to attempt cannulation of other arteries. The left carotid arterial flow was monitored by an overlying doppler probe and frequent palpations of the carotid pulses were carried out. Estimations of blood pressure were determined by the sound of a doppler flowmeter placed securely over the left carotid artery and by palpation of the carotid pulses on a frequent and regular basis to detect changes in the intensity of the sound and strength of pulsations.

During spontaneous respiration under deep oxygen and halothane anaesthesia, several attempts with various straight laryngoscope blades, positioning of the head and with posteriorly directed pressure on the larynx were required before an uncuffed 4.5 mm oral endotracheal tube with stylet was inserted. Subsequent examination of the intubated glottis by direct laryngoscopy revealed an acute inversion of the epiglottis which was displaced to the right and a dislocated left arytenoid cartilage. Gentle manipulation with Magill forceps successfully reduced the epiglottis and arytenoid to their anatomical positions.

Anaesthesia was maintained with nitrous oxide, oxygen and halothane and ventilation was mechanically controlled. During the three-and-a-half-hour procedure 400 ml of D5W-0.2 per cent NS was administered for maintenance and replacement of fluid deficit.

Blood samples were drawn by retrograde aspiration of the IV cannula in the external jugular vein. Venous blood gases revealed a normal pH, normocarbia and a mild metabolic acidosis, and a haemoglobin of 90 $g \cdot L^{-1}$. Previous bloodwork showed a haemoglobin of 107 $g \cdot L^{-1}$. Additional blood was drawn during the case for iron studies by the Department of Haematology. The patient received 10 mEq $NaHCO_3$ to counter his lowered bicarbonate level which was presumed to be secondary to starvation ketosis and also received dexamethasone IV because of the possibility of glottic oedema due to intubation difficulties at induction.

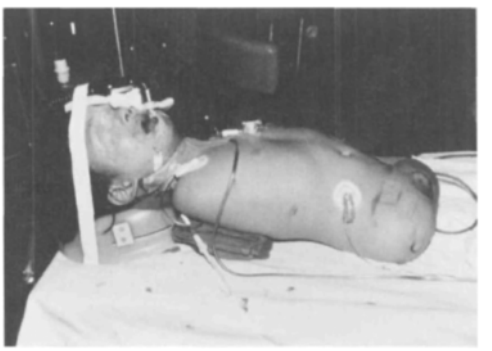

FIGURE 1 Anaesthetized four-year-old patient with amelia.

The patient awakened and was extubated uneventfully at the termination of his dental work. After a three-hour stay in the Recovery Room he was discharged home.

Discussion

Amelia or ectromelia denotes a complete absence and meromelia a partial absence of limb or limbs. Phocomelia ("seal flippers") and hemimelia ("half limb") are older descriptive terms abandoned by current nomenclature.

Limb buds begin developing towards the end of the fourth gestational week, those of the legs appearing a few days after those of the arms. By the seventh gestational week these progressively elongating appendages, consisting of mesenchyme covered by a layer of ectoderm have formed toes and fingers and have rotated into anatomical orientation. Further development involves continued growth into limbs of vessels and nerves. Complete absence of a limb is attributed to arrest of primary limb bud formation and partial absence of a limb is attributed to developmental disturbance at a later stage.[1]

Aetiological factors cited in limb deficiencies are genetic and chromosomal, mechanical intrauterine disruptions and influences such as oligohydramnios, and most often teratogenic drugs and chemicals.[2]

O'Rahilly and Frantz[3] estimated by extrapolation a prevalance of amelia of 1 in 3.5 million. Heinomen *et al.*[4] analyzed data from a prospective study of 50,282 pregnancies from 1958 to 1965 and counted 32 births with variable degrees of limb deficiency. There was a significant percentage of central nervous system, cardiovascular and musculo-skeletal anomalies as well as identifiable syndromes in this group. Amelic patients may have elevated baseline temperatures since without limbs they have less surface area through which they can eliminate heat. Ivankovitch *et al.*[5] in a case report describe a 13-year-old amelic girl with progressive scoliosis who underwent insertion of Harrington rods and spinal fusion with autologous iliac grafts. A right internal jugular CVP line was inserted two days preoperatively and BP was monitored by superficial temporal artery canulation inserted by cutdown under incremental IV ketamine prior to intubation. Surgery was performed in the prone position and deliberate hypotension was employed. Surgery and recovery were uneventful.

Thiagarajah *et al.*[6] in a case report describe non-invasive blood pressure monitoring in a vasculopathic 68-year-old woman undergoing revision of a right axillo-bifemoral graft. Poorly perfused limbs made conventional blood pressure monitoring impossible and areterial cannulation seemed technically too difficult. Blood pressure was instead recorded by placing a doppler probe over the superficial temporal artery and a Kidde tourniquet around the head. In their discussion the authors cite comparison of blood pressures measured this way and by brachial cuff in a group of volunteers and noted a fairly good agreement in measurements between the two techniques.

Ideally, patients who are ASA physical status III or greater should not undergo general anaesthesia in the day care setting. As a rule all patients who present with problems with major anaesthetic implications are seen in advance in our Anaesthesia Clinic and generally receive their general anaesthetic in the main ORs of the hospital. This patient presented with difficulties which normally would have precluded anaesthesia on an outpatient basis, but circumstances dictated the course chosen.

Intravascular cannulation to obtain direct blood pressure measurement and to provide access for determination of arterial blood gases would be ideal. After unsuccessful cannulation of the iliac vessels, it was decided that carotid and/or temporal artery cannulation was not warranted. Sampling from the external jugular cannula provided blood gas, acid base balance, and haematological measurements.

This patient presented with a difficult airway which may be one of the associated congenital anomalies with amelia. The induction of anaesthesia in patients with upper airway deformities which may make intubation difficult should always be carried out by awake intubation using topical anaesthesia or by the inhalation method. No muscle relaxants should be used. Indeed this patient proved to be very difficult to intubate due to the anterio superior placement of the larynx. In addition, this case provides an excellent example to compel all anaesthetists to directly examine the airway following a difficult intubation to assure that the laryngeal structures are in their correct anatomical position. Using the oral route for dental surgery increases the hazard of obstructing the airway and/or accidental extubation. However, in this case, the oral route was mandatory because the anatomical difficulties made it necessary to use an oral tube and stylet.

This patient was subsequently admitted to hospital seven months later and underwent bilateral orchidopexies. The anaesthetic management was essentially the same and was uneventful.

Summary

A four-year-old boy was given a general anaesthetic for dental caries repair. The absence of limbs made it difficult to monitor his CVS system and also created problems securing IV and arterial access. He also presented with a moderately difficult intubation and his airway had to be shared with the dentist.

Patients with amelia are very rare but the surgery they require may be major. They may present with anomalies of major organ systems and present the anaesthetist innumerable challenges in the anaesthetic management.

References

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Résumé

Un garçon âgé de quatre ans né sans membres (amelie) s'est présenté pour restaurations dentaires sous anesthésie générale comme patient externe. Après l'administration d'atropine par voie intramusculaire l'anesthésie a été induite avec l'halothane, oxygène et protoxyde d'azote au masque. Par la suite, une voie intraveineuse a été assurée par la cathétérisation de la veine jugulaire externe. A cause de sa petite bouche, d'un hypognatisme, d'un palais arqué et d'un larynx situé dans une position antéro-supérieure, l'intubation orale sous anesthésie générale profonde et ventilation spontanée était difficile. L'épiglotte était inversée. Lors d'une inspection subséquente au laryngoscope et après intubation, fût réduite mécaniquement à sa position anatomique. Malgré l'incapacité de surveiller précisément la pression artérielle, la période per-opératoire s'est accomplie sans incident. En période post-opératoire le patient était extubé et pouvait retourner à la maison le même jour.