Intraoperative death during Caesarian section in a patient with sickle-cell trait

The Anaesthesia Advisory Committee
to the Chief Coroner of Ontario*

The case of a woman with sickle cell trait who sustained a cardiac arrest and died during a Caesarian section under general anaesthesia is reported. Because the common causes of intraoperative hypoxia and shock were ruled out in this case, we believe that death was due to severe concealed aorto-caval compression.

After delivery, the release of a large volume of hypoxaemic, acidotic blood with sickled cells could cause cardiac depression and arrest. The fact that the patient's mucous membranes were pink and she was haemodynamically stable while her uterus was cyanotic prior to delivery provides some positive evidence for this hypothesis.

We emphasize that while complications secondary to sickle cell trait during general anaesthesia are very rare, they can occur. We discuss methods of monitoring such patients.

Haemoglobin S is a haemoglobin variant (valine replaces glutamic acid in the sixth position of the β-chain) which forms a gel-like structure when deoxygenated. This causes reversible and eventually irreversible changes in red cell morphology.[1]

Patients who are homozygous for HbS (sickle cell disease – SCD) have significant illnesses and shortened life expectancy, but those who are heterozygous (sickle cell trait – SCT) have lower amounts of haemoglobin S (25–45 per cent of total haemoglobin) and are usually thought to be free of *in vivo* sickling and to be basically

Key words

COMPLICATIONS: cardiac arrest, death; SURGERY: Caesarian section; BLOOD: sickle cell trait; SHOCK: aorto-caval compression.

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normal except for an increased incidence of haematuria,[2] possibly because of microinfarcts in the renal papillae.[3]

In keeping with this view of SCT, a review of anaesthetic management concluded: "Patients with sickle cell trait are at no greater risk during anaesthesia and surgery than patients with normal haemoglobin."[4] However, further correspondence emphasized that there is "an unwarranted assumption in the summary that the anaesthetist can always guarantee adequate oxygenation."[5] Although SCT is not uncommon (eight per cent of blacks[6] and more rarely in East Indians and individuals of eastern Mediterranean origin) there are few reports of intraoperative and postoperative deaths in patients with SCT[7] and at least one of these[8] was subsequently found to have sickle cell – HbC disease[9] (a combination of sickle cell trait with HbC which is a much more serious condition). Many others were found to be unrelated to the anaesthetic[4].

Therefore we wish to present a case of intraoperative death during Caesarian section under general anaesthetic which was the subject of a recent coroner's inquest. We believe the death was secondary to severe aortocaval compression in a patient with SCT.

Case report

A previously healthy 28-year-old pregnant black woman was admitted close to term in early labour with ruptured membranes. She had had four uneventful general anaesthetics in the past including one for a Caesarian section for cephalopelvic disproportion three years before. At that time, she had received thiopentone, succinylcholine, pancuronium, Innovar, fentanyl and N_2O during the anaesthetic. On the current admission, her cervix was long and uneffaced so an emergency repeat Caesarian section was planned. The patient was known to have sickle cell trait but this information was not noted by the operating room staff.

Two hours after admission the patient was taken to the operating room, where she was examined by the anaesthetist. The following data were recorded: height 163 cm, weight 74 kg, haemoglobin 129 g·L^{-1}, pulse 88/min., blood pressure 110/70 mmHg, temperature 36.7° C, fetal heart rate 140–145/min. Chest examination was normal.

There was no peripheral oedema. The patient had eaten last five hours preoperatively. She requested general anaesthesia.

A Boyle anaesthesia machine with a 30–70 per cent proportioner mixing valve was used, along with a Medishield floating bellows ventilator incorporating an automatic disconnect alarm. Monitoring included electrocardiogram, temperature and pulse monitors, and a blood pressure cuff. Testimony given at the Inquest by everyone present at the operation was in general agreement on the course of events.

The patient was tilted slightly to the left with one folded OR sheet under her right hip, and given 1 ml of Innovar IV. After five minutes of breathing 100 per cent O_2 she was induced with fentanyl 50 μg, thiopentone 225 mg and succinylcholine 50 mg, with the use of cricoid pressure during the induction sequence. She was intubated easily with a 8 mm cuffed ETT and both sides of the chest were auscultated. Normal breath sounds were heard. She was then given 4 mg of pancuronium and ventilated with 60 per cent N_2O:40 per cent O_2 (minute volume 8 L). Blood pressure was taken six times prior to delivery and varied between 130 and 110 systolic. The pulse rose from 90 to 110 bpm prior to delivery. Three minutes before the delivery the patient was placed on 100 per cent O_2 and given naloxone 0.2 mg IV. Delivery occurred 23 minutes after the start of preoxygenation and 33 minutes after the patient was placed on the operating room table. Skin incision to delivery time was nine minutes. Immediately following delivery, 60 per cent N_2O was reintroduced and the patient was given 250 μg of fentanyl and five units of syntocinon IV.

Although the skin and abdominal wall incision was unremarkable, the surgeon noted the uterus was dark when he opened the abdomen and dark blood escaped from the uterus when it was incised. When informed of this, the anaesthetist rechecked the patient's blood pressure and pulse. These were unchanged. He checked her buccal mucus membranes and they were pink. The baby was born asphyxiated (Apgar score 2 at one minute) and the anaesthetist took the baby to resuscitate it.

With immediate intubation and ventilation with 100 per cent O_2 and IV naloxone (0.04 mg) the baby was pink and vigorous in one to two minutes (Apgar score ten at six minutes). While the anaesthetist was resuscitating the baby, he noted the maternal pulse rate dropping by listening to the ECG audio output and asked the circulating nurse to check the patient's blood pressure. It was unobtainable. The anaesthetist was able to extubate the baby and return to the mother immediately. The surgeon reported the aortic pulse was weak and slow. The patient was ventilated manually with 100 per cent O_2 and chest movement was checked. Despite atropine 1.2 mg, and

two 20 mg doses of ephedrine IV, the aortic pulse became impalpable and CPR was begun. During resuscitation, the cardiac arrest team and a second anaesthetist arrived to assist. Despite multiple doses of epinephrine and sodium bicarbonate, a fluid bolus, $CaCl_2$, and an isoproterenol drip, the patient progressed from the original sinus bradycardia with electromechanical dissociation to an agonal rhythm to asystole and was pronounced dead.

A chest x-ray done on the operating table showed the endotracheal tube in correct position. Blood gases drawn ten minutes after CPR started showed pH 7.13, pCO_2 74 mmHg (10 kPa) and pO_2 16.3 mmHg (2.2 kPa). Blood was taken from the femoral area and could have been venous. Autopsy findings revealed only congestion of the viscera (lungs, liver, spleen, kidneys, and pancreas) with widespread sickling. Haemoglobin electrophoresis revealed 56 per cent HbA and 40 per cent HbS. There was no coronary or valvular heart disease, no cardiomyopathy, no emboli (of air, amniotic fluid or thrombus), no cardiac tamponade and no hidden blood loss. The stomach contained partially digested food. There were no particles or other signs of aspiration in the airways. The anaesthetic machine was checked immediately following the incident by a second anaesthetist and a respiratory technologist. It delivered appropriate O_2 concentrations. It was impounded and checked twice later on (by the manufacturer and by the Medical Devices Branch of Health and Welfare, Canada) and no defects which would significantly affect its function were discovered.

Discussion

The circumstances of this death, as established during the Inquest, rule out most of the common causes of intraoperative hypoxia or shock (e.g., hypoxic gas mixtures, aspiration of gastric contents, circuit disconnection, hypoventilation, misplaced endotracheal tube, cardiomyopathy, myocardial infarction, cardiac tamponade, sepsis, bleeding or embolus). This leaves only a few uncommon possibilities. Anaphylaxis to any of the anaesthetic drugs is unlikely because they were given almost twenty minutes prior to the haemodynamic deterioration. Sympathetic overactivity and arrest following naloxone is equally unlikely because the deterioration only occurred after injection of more fentanyl and reinstitution of nitrous oxide. Also, the patient never had severe tachycardia or ventricular ectopics which would be expected in this setting. Certainly the anaesthetic technique used was unusual. It does not follow the recommended pattern of 50 per cent N_2O:O_2 plus a low dose of an inhalation agent prior to delivery. The technique used could lead to maternal awareness, but does not account for the findings in this case.

This leaves as the only possibility that there had been a

severe form of concealed aortocaval compression[10] while the patient was in the supine position (she was not tilted very much – probably less than 3 cm with the folded OR sheet). This would explain the congested, cyanotic uterus prior to delivery while the upper body circulation was still stable and the mucus membranes pink.

Following delivery, a large proportion of the patient's total blood volume, which may have been stagnant for more than 30 minutes, would be released suddenly. This blood would be acidotic, with a high potassium concentration and could contain a significant proportion of sickled cells.

When this bolus reached the heart it could cause cardiac depression and bradycardia and a vicious cycle of further hypoxia and stasis would be started. Once this was well under way, the presence of the sickle cell trait would make resuscitation difficult or impossible. The anaesthetic chart for the first Caesarian section did not mention tilting but evidence given at the Inquest was that tilting was a routine practice at that time. However, differences in the size and shape of the uterus in the two pregnancies could have led to differences in aortocaval compression.

Recent literature on the sickle cell trait has emphasized the fact that complications may be very rare but they do occur. Diggs, in his extensive review[7] discusses several areas of concern. Patients with SCT have had splenic infarcts when exposed to altitude (greater than 3000 m) and *in vivo* sickling has been discovered in people with SCT at this altitude. The incidence of sudden death during exercise is higher in young men with SCT. In one study of military training, all four sudden deaths occurred in individuals with SCT.[11] Therefore, because of these risks Diggs[12] concludes that military personnel with sickle cell trait should be excluded from training and assignment as aircrew or divers, and should be excluded from special operations that require maximum stamina and exertion.

Similarly, anaesthetists should recognize that while problems with patients with sickle cell trait may be rare, they are not non-existent. If patients with SCT become severely hypoxic, acidotic or shocked from any cause, they may be difficult to resuscitate. Therefore every effort should be made to determine the sickle cell status of patients in the high risk groups preoperatively, by history and routine sickle cell testing.

All patients with SCT undergoing general anaesthesia should be monitored to assure constant adequate tissue oxygenation. The pulse oximeter or transcutaneous pO_2 monitors represent convenient non-invasive methods. Special precautions must be taken to avoid local ischaemia, stagnation or cooling. No tourniquets should be used. Special precautions, including exchange transfusion, may be necessary for open heart surgery. Placing the pulse oximeter probe on the patient's toe during Caesarian

section should monitor the adequacy of the patient's tilt to the left. The pulse oximeter would detect any significant hidden aortocaval compression and thus prevent local stagnation and hypoxia below the level of the uterus.

An important reminder from this case is that a second physician trained in infant resuscitation should be present at all Caesarian sections. This allows the anaesthetist to concentrate solely on the mother and to avoid any need to deal with two critically ill patients simultaneously.

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Résumé

*On rapporte l'histoire d'une femme porteuse d'une drépano-
cytose latente ayant subi une césarienne sous anesthésie
générale au cours de laquelle elle a présenté un arrêt cardiaque
suivi d'un décès. Parce que les causes communes d'hypoxie
intraopératoire et l'état de choc ont été éliminés dans ce cas, on
pense que le décès était dû à une compression sévère et non
détectée de l'aorte ainsi que de la veine cave.*

*Après la délivrance, la libération d'un grand volume de sang
hypoxémique, acidotique avec des cellules falsiformes a pu
provoquer une dépression myocardique et un arrêt. Le fait que
les membranes muqueuses de la patiente étaient rosacées et
qu'elle présentait un état hémodynamique stable alors que son
utérus était cyanotique avant la délivrance, fournissait une
preuve positive de cette hypothèse.*

*On suggère que même si les complications secondaires à la
drépanocytose lors de l'anesthésie générale sont très rares,
elles peuvent cependant survenir. On discute des méthodes de
surveillance dans tels cas.*