Clinical Reports

Cesarean section in a mother with uncorrected congenital coronary to pulmonary artery fistula

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Purpose: We report a case of a 33 yr old woman with pulmonary hypertension secondary to uncorrected right coronary artery to pulmonary artery fistula who underwent two successful operative deliveries under general anesthesia.

Clinical Features: This woman underwent an emergency Caesarean section at 32 wk gestation because she presented in NYHA Class IV, heart failure and premature labour. She did not have antenatal follow-up. For her second pregnancy, she was managed from the first trimester of pregnancy by the cardiologist, obstetrician and anesthesiologist. She received oral furosemide and digoxin from eight weeks gestation. Pregnancy was managed to term before she progressed to NYHA Class IV and cardiac failure at 37 wk gestation. She had a Caesarean section under general anesthesia. She received rapid sequence induction of anesthesia and tracheal intubation with 0.1 mg·kg⁻¹ etomidate, 2 mg·kg⁻¹ succinylcholine and maintenance with nitrous oxide 50% in oxygen, isoflurane 1% and 0.1 mg·kg⁻¹ vecuronium. Fentanyl, 2 µg·kg⁻¹, helped to obtund the hypertensive response to intubation. Analgesia was provided with 1 mg·kg⁻¹ morphine. Glyceryl trinitrate infusion, 10-30 µg·min⁻¹, was used in addition to the anti-heart failure therapy. End-tidal capnography, electrocardiogram, pulse oximetry, continuous arterial blood pressure and pulmonary arterial catheter provided hemodynamic monitoring. The lungs were mechanically ventilated for 24 hr postoperatively. She received anti-heart failure therapy which she continued after discharge. She was NYHA class II upon discharge. She defaulted from further follow-up.

Conclusion: Although the literature advocates, in this situation, controlled vaginal delivery utilising epidural analgesia, we describe the successful outcome for operative delivery under general anesthesia in a patient with secondary pulmonary hypertension and heart failure.

Objectif : Rapporter le cas d'une femme de 33 ans atteinte d'hypertension pulmonaire, secondaire à une fistule non corrigée entre l'artère coronaire droite et l'artère pulmonaire, qui a subi avec succès deux césariennes sous anesthésie générale.

Éléments cliniques : Cette patiente, sans suivi prénatal, a subi une césarienne d'urgence à 32 sem de grossesse parce qu'elle s'est présentée souffrant de défaillance cardiaque de classe IV NYHA (New York Heart Association), et en travail prématuré. Vue dès le premier trimestre, pendant sa seconde grossesse, par le cardiologue, l'obstétricien et l'anesthésiologiste, elle a reçu du furosémide peroral et de la digoxine à partir de la huitième semaine. La patiente a pu mener sa grossesse à terme avant de présenter la Classe IV NYHA et la défaillance cardiaque à 37 sem. Une césarienne sous anesthésie générale a été réalisée : l'induction de séquence rapide et l'intubation endotrachéale avec 0,1 mg·kg⁻¹ d'étomidate et 2 mg·kg⁻¹ de succinylcholine, le maintien avec un mélange de protoxyde d'azote et d'oxygène à 50 %, de l'isoflurane 1 % et 0,1 mg·kg⁻¹ de vécuronium. La réduction de l'hypertension liée à l'intubation a été assurée par 2 µg·kg⁻¹ de fentanyl et l'analgésie par 1 mg·kg⁻¹ de morphine. Une perfusion de trinitrate de glycérile, 10-30 µg·min⁻¹, a été ajoutée au traitement anti-défaillance cardiaque. Le monitoring hémodynamique a été réalisé par la capnographie de fin d'expiration, l'oxymétrie pulsée, la surveillance continue de la tension artérielle et un cathéter de l'artère pulmonaire. La ventilation pulmonaire mécanique postopératoire s'est poursuivie pendant 24 h. La patiente a reçu un traitement anti-défaillance cardiaque qu'elle a continué après son départ de l'hôpital. Elle était alors à la classe II NYHA. Elle ne s'est pas présentée à la visite de contrôle.

Conclusion : Bien que les recherches conseillent un accouchement vaginal contrôlé et une analgésie péridurale dans ce genre de situation, nous citons ici l'heureux résultat d'une césarienne sous anesthésie générale chez une patiente qui présentait une hypertension pulmonaire secondaire et une défaillance cardiaque.

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IN congenital coronary arteriovenous fistula there is a communication between a coronary artery and the lumen of any one of the four cardiac chambers, the coronary sinus, pulmonary artery, pulmonary veins or superior vena cava.¹ We report the successful perioperative management of a parturient with an uncorrected right coronary artery to pulmonary artery fistula who underwent two operative deliveries.

Case report

This 33 yr old Chinese woman was told, since childhood, that she had a heart murmur but defaulted on investigations and was managed by a traditional healer. Her first wanted pregnancy terminated in a miscarriage in 1989. Physical examination at that time revealed a febrile patient with tachycardia, a displaced apex beat and pansystolic murmur, grade 3/6. There was no sign of heart failure or infective endocarditis. Electrocardiogram showed sinus tachycardia, P pulmonale, left axis deviation and left ventricular hypertrophy with strain pattern. The chest radiograph showed prominent pulmonary arteries and cardiomegaly and a 2D echocardiogram showed a left to right shunt into the main pulmonary artery. The right coronary artery, pulmonary artery, left atrium, left

ventricle and the right atrium were enlarged. The diagnosis was that of a right coronary artery to pulmonary artery fistula. The febrile state was thought to be due to a septic abortion. Cardiac catheterisation was planned but she defaulted investigation.

Four years later, she presented in cardiac failure and premature labour at 32 wk gestation. Clinically, she was New York Heart Association (NYHA) Class IV. Oxygen therapy, intravenous furosemide, digoxin and potassium supplement were started. A 2D echocardiogram confirmed the presence of a fistula between the pulmonary artery and the right coronary artery. There was pulmonary hypertension (mean PAP was 60mmHg), left ventricular hypertrophy, dilated left atrium and ventricle. The ventricular ejection fraction was 67%. A pulmonary artery catheter revealed the hemodynamic data shown in Table I. She did not respond to the anti-heart failure therapy. Caesarean section was performed in view of her cardiac failure and possible hemodynamic instability. End-tidal capnography, electrocardiogram, pulse oximetry, continuous arterial blood pressure and pulmonary arterial catheter provided hemodynamic monitoring. She underwent uneventful general anesthesia employing rapid sequence induction using 0.1 mg·kg⁻¹ etomidate, 2 mg·kg⁻¹ succinylcholine and maintenance with nitrous oxide 50% in

TABLE I Hemodynamic data during first pregnancy

	<i>Pre-operative data</i>	<i>Post-induction data</i>	<i>4 hr postoperative data</i>
CO (L·min ⁻¹)	6.4	6.0	10.2
CVP (mmHg)	12	10	4
MAP (mmHg)	89	81	75
HR (bpm)	101	105	110
mPAP (mmHg)	75	78	71
PAWP (mmHg)	55	48	16
PVR (dyne·sec·cm ⁻⁵)	201	203	188
SVR (dyne·sec·cm ⁻⁵)	1029	1009	1050

CO = cardiac output; CVP = central venous pressure; MAP = mean arterial pressure HR = heart rate; mPAP = mean pulmonary arterial pressure; PAWP = pulmonary arterial wedge pressure; PVR = pulmonary vascular resistance; SVR = systemic vascular resistance

TABLE II Hemodynamic data during second pregnancy

	<i>Pre-operative data</i>	<i>Post-induction data</i>	<i>4 hr postoperative data</i>
CO (L·min ⁻¹)	12	11	8
CVP (mmHg)	10	9	8
MAP (mmHg)	93	89	85
HR (bpm)	100	109	122
mPAP (mmHg)	72	74	75
PAWP (mmHg)	28	25	18
PVR (dyne·sec·cm ⁻⁵)	270	280	362
SVR (dyne·sec·cm ⁻⁵)	900	869	1020

CO = cardiac output; CVP = central venous pressure; MAP = mean arterial pressure HR = heart rate; mPAP = mean pulmonary arterial pressure; PAWP = pulmonary arterial wedge pressure; PVR = pulmonary vascular resistance; SVR = systemic vascular resistance

oxygen, isoflurane 1% and 0.1 mg·kg⁻¹ vecuronium. Fentanyl, 2 µg·kg⁻¹, helped to obtund the hypertensive response to intubation. Analgesia was provided with 1 mg·kg⁻¹ morphine. Intravenous ampicillin, gentamycin were given for endocarditis prophylaxis. The trachea was extubated after the surgery and she was managed in the surgical intensive care. Glyceryl trinitrate infusion, 0.6-1.8 mg·hr⁻¹, was used. She refused tubal ligation. She was discharged with 40 mg furosemide q12h, 1.2 gm potassium chloride q12h and 0.625 mg digoxin q24h *po* and was then functionally NYHA Class II. She again defaulted cardiac follow up.

In this present pregnancy she was referred to the obstetrician at eight weeks gestation and was managed together with the cardiologist. She was clinically NYHA Class I. She was restarted on oral anti-heart failure therapy. However, she progressed to NYHA Class IV by 37 wk gestation despite therapy. In view of her cardiac failure, history of previous successful operative delivery under similar conditions, Caesarean section was planned. She also agreed to concomitant tubal ligation. The anesthesiologist electively inserted a pulmonary artery catheter which revealed the hemodynamic data shown in Table II. She received a similar general anesthetic technique to that of her previous Caesarean section. The lungs were mechanically ventilated post operatively to avoid hypoxemia and hypercarbia. The trachea was extubated 24 hr later and she remained in the surgical intensive care for four days to optimise her cardiac status. She responded well to 80 mg furosemide q8h *iv*, fluid restriction 1 mg·hr⁻¹ morphine and 0.6 - 1.8 mg·hr⁻¹ glyceryl trinitrate infusion. The pulmonary artery catheter was removed 48 hr later. She was NYHA class II on discharge. She again defaulted cardiac follow-up.

Discussion

We report a case of congestive cardiac failure and pulmonary hypertension secondary to uncorrected congenital right coronary artery to pulmonary artery fistula who successfully underwent two operative deliveries under general anesthesia.

Heart disease during pregnancy occurs in 0.4-4.1% of patients and is the leading non-obstetric cause of maternal mortality ranging from 0.4% among patients in NYHA Class I/II to 6-8% in NYHA Class III/IV patients.² Cardiac decompensation occurs most commonly at the time of maximum hemodynamic stress in the third trimester of pregnancy, during labour and delivery and in the immediate postpartum period.

We postulate that the pulmonary hypertension in our patient was secondary to long term increased blood flow from the right coronary artery to the pulmonary artery. Fortunately she did not have Eisenmenger's syndrome.

Anesthesia for Caesarean section may be provided by either general anesthesia or epidural technique. General anesthesia provides better control of ventilation and oxygenation.³ Epidural anesthesia has the risk of producing too high a sympathetic blockade resulting in severe hypotension.³ Despite these risks epidural anesthesia has been used satisfactorily for Caesarean section and particularly for vaginal delivery.^{4,5,6}

The usefulness of a pulmonary artery catheter is controversial.⁷ However, it can provide information about left ventricular function and pulmonary vascular changes.⁸ We chose to insert a pulmonary artery catheter because the data obtained were relevant to the patient's management.

The natural history of coronary arteriovenous fistula is not precisely known. Daniel and colleagues found that congestive heart failure occurred either in infancy or after 20 yr of age with a peak incidence in the fifth and sixth decades.⁹ Our patient developed cardiac failure because of pregnancy induced hemodynamic changes and cardiac volume overload.¹⁰ Other complications like fistula dissection and rupture are rare.¹⁰ Surgery is potentially curative. Sadly, this patient repeatedly defaulted cardiac follow-up.

Conclusion

Congenital coronary arteriovenous fistula is uncommon. This patient developed high flow congestive cardiac failure and pulmonary hypertension due to pregnancy induced hemodynamic changes. She did not have Eisenmenger syndrome. We report her successful maternal and fetal outcome following Caesarean section under general anesthesia with cardiovascular monitoring. Multidisciplinary team approach to management contributed to the successful outcome.

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