Transient iatrogenic heart block following fetal intracardiac transfusion for severe twin anaemia-polycythaemia sequence

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**Abstract:**
Background: Intrauterine blood transfusion is an important treatment of fetal anaemia. Although, the standard access to the fetal vasculature for transfusion is the umbilical vein, the intracardiac route is used when fetal or placental positions make other accesses technically challenging. Intrauterine, intracardiac blood transfusion is associated with complications including haemopericardium, damage to cardiac tissues and fetal bradycardia.

Highlights of the present report: We report a case of monochorionic twins with twin anaemia-polycythaemia sequence. Intracardiac, intrauterine blood transfusion of the donor twin was complicated by haemopericardium and sustained bradycardia which necessitated delivery by emergency cesarean section. Postnatally, the bradycardia was sustained and was diagnosed electrocardiographically as heart block, which spontaneously reversed on the second day after birth. The management of heart block in the neonatal period is discussed.

Conclusion: Fetal intracardiac intrauterine blood transfusion can be associated with transient congenital heart block.
Re: CRPM.2016.0004

Transient iatrogenic heart block following fetal intracardiac transfusion for severe twin anaemia-polycythaemia sequence

Case Reports in Perinatal Medicine

Dear Professor Dudenhausen,

We would like to thank the reviewer for the constructive comments that have helped us improve the quality of our manuscript. Please find appended our detailed response to the points raised by the reviewer. We have revised the manuscript addressing the individual concerns raised.

Kind regards

On behalf of the authors

Theodore Dassios, MD, PhD, FRCPCH
RESPONSE TO REVIEWERS COMMENTS

Reviewer: 1, Comments to the Author

Reviewing this case report caused a bit of a dilemma; the first reaction reading this paper, and I am sure many in the field will feel the same, is that there is no reason to choose for the intracardiac route which everyone knows is more dangerous in any fetus at 29 weeks. The umbilical vein can be accessed at the plental insertion site, the fetal liver or a free loop, all safer. The authors however address this limitation, and therefore the focus of the importance of the paper should be the transient heart block, which indeed is new and could have some importance to know about for the fetal therapy community. Otherwise well written and clear, although a few more details on the prenatal diagnosis of TAPS would be nice.

Response: We would like to thank the reviewer for the kind comments and assessment of our work. We have expanded our introduction on the prenatal diagnosis of TAPS to include the following: “TAPS can be diagnosed antenatally or postnatally. The absence of oligohydramnios and polyhydramnios on antenatal ultrasound is a prerequisite for the diagnosis of TAPS, since these signs are pathognomonic for the twin oligopolyhydramnios sequence. Prenatal diagnosis of TAPS is based on Doppler ultrasound abnormalities showing decreased peak systolic velocity in the middle cerebral artery (MCA-PSV) in the recipient twin (suggestive of fetal polycythaemia) and increased MCA-PSV in the donor twin (suggestive of fetal anaemia).”
ABSTRACT

Background: Intrauterine blood transfusion is an important treatment of fetal anaemia. Although, the standard access to the fetal vasculature for transfusion is the umbilical vein, the intracardiac route is used when fetal or placental positions make other accesses technically challenging. Intrauterine, intracardiac blood transfusion is associated with complications including haemopericardium, damage to cardiac tissues and fetal bradycardia.

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Keywords: fetal intrauterine blood transfusion, congenital heart block, cardiocentesis

Word count: 1052
INTRODUCTION

Chronic feto-fetal transfusion due to placental vascular anastomoses in monochorionic twin pregnancies may culminate in twin anaemia-polycythaemia sequence (TAPS). The ensuing communications result in transfusion of blood from the donor to the recipient twin gradually leading to a severely anaemic donor twin and a severely polycythaemic recipient co-twin [1]. TAPS can be diagnosed antenatally or postnatally. The absence of oligohydramnios and polyhydramnios on antenatal ultrasound is a prerequisite for the diagnosis of TAPS, since these signs are pathognomonic for the twin-oligopolyhydramnios sequence. Prenatal diagnosis of TAPS is based on Doppler ultrasound abnormalities showing decreased peak systolic velocity in the middle cerebral artery (MCA-PSV) in the recipient twin (suggestive of fetal polycythaemia) and increased MCA-PSV in the donor twin (suggestive of fetal anaemia) [2].

Laser coagulation of the small anastomoses is the only treatment of TAPS but this may be technically challenging, especially at advanced gestation. Intrauterine transfusion of the donor twin with partial exchange transfusion of the recipient twin is an alternative option with a view to prolonging the pregnancy, as this may temporarily alleviate the severity of the condition of both twins[3]. The technique for intravascular fetal blood transfusion has been described in the treatment of severe alloimmunisation in rhesus disease[4]. The umbilical vessels are commonly preferred to access the fetal circulation [5] but the intracardiac route is used when access to the umbilical vessels is not feasible due to placental or fetal position. The intracardiac route has been associated with fetal complications such as haemopericardium, asystole and severe fetal bradycardia[6]. We now novelly report a fetus/infant who suffered heart block as a consequence of the procedure and discuss the management of heart block presenting in the perinatal period.
PRESENTATION OF THE CASE

The mother had a spontaneously conceived monochorionic, diamniotic twin pregnancy. Ultrasound scan at 29 weeks of gestation was suggestive of TAPS. The following management options were discussed with the parents: (a) expectant management with risk of fetal demise of one or both twins; (b) iatrogenic preterm delivery with significant prematurity-related morbidity; (c) endoscopic laser ablation of the placental anastomoses with substantial failure rate due to advanced gestation and (d) intrauterine blood transfusion of the anaemic (donor) twin with partial exchange transfusion of the polycythaemic (recipient) co-twin. Following counselling, the parents opted for the latter option. In view of placental and fetal position, the intracardiac route to transfuse the donor was deemed the most appropriate.

Normal sinus rhythm prior to the procedure was demonstrated for both twins. Access to the donor fetal heart was obtained via ultrasound-guided insertion of a spinal needle into the right ventricle. Fetal haemoglobin prior to the transfusion was 3.8g/dl. Following intracardiac removal of 40ml of fetal blood and intracardiac transfusion of 70ml of ORh negative donor blood, fetal haemoglobin of the donor twin increased to 9.5g/dl. The procedure however was complicated by haemopericardium and fetal bradycardia and the twins were delivered via emergency caesarean section.

The donor female twin infant was born with a heart rate of less than 60 bpm and no respiratory effort. Endotracheal intubation was undertaken at two minutes of age. A blood transfusion at 20 ml/kg and normal saline at 10 ml/kg were administered. On admission to the neonatal unit, the heart rate was 70-80 bpm and haemoglobin was 13.8 g/dl. A chest radiograph taken at two hours of age revealed a left-sided pneumothorax and pneumopericardium (Figure 1) which were evacuated with no effect on the heart rate.
Echocardiography at five hours of age revealed a structurally normal heart; twelve-lead
electrocardiography revealed complete heart block with ventricular rate of 70-78bpm (Figure
2). Ventricular heart rate fluctuated between 60 and 80bpm until 34 hours of age, when
atrioventricular (AV) conduction spontaneously recovered at a rate of 120-140bpm. The chest
drains were removed on day four after birth and the infant was successfully extubated on day
five after birth. She was discharged well to the local hospital on day 19. No evidence of
intracranial pathology was demonstrated on serial cranial ultrasound examinations.

The recipient twin sibling underwent a dilutional exchange transfusion on admission and was
subsequently discharged to the local hospital on day 19.

DISCUSSION

This report highlights a novel association of heart block and fetal intracardiac blood
transfusion. Percutaneous entry to the fetal heart represents an alternative route by which the
fetal circulation can be accessed if the umbilical vein is not accessible[7]. Comparative
observational data suggest that the intracardiac route carries high procedure-associated risk
and 33% mortality[6]. Of note, the currently accepted complication rate of 6.8% of
intracardiac intrauterine transfusion is derived from twenty-five cases in which the procedure
was used due to Rhesus isoimmunisation in severely compromised fetuses[6].

In this case report the normal fetal heart rate before the procedure, the duration and resolution
of the heart block and return to AV conduction suggest that the conduction system might
have been injured during the cardiocentesis. Traumatic iatrogenic heart block is transient and
self-resolving with supportive treatment. Traumatic AV block occurs in 2% of all cardiac
operations and is associated with 7% mortality within the first 30 postoperative days owing to
haemodynamic instability[8]. Recovery of AV conduction occurs in 55-100% of cases; 95% of the patients who regain AV conduction do so by postoperative day 9[8]. Cases that fail to regain AV conduction require permanent pacemaker implantation.

Traumatic heart block differs to congenital heart block (CHB) secondary to autoimmune disease and structural heart disease in that it follows a course that is more benign and of shorter duration. CHB is a rare disorder, defined as the conduction system disease of any form diagnosed within the first 28 days of life[9]. It is often related to maternal autoimmune disease and congenital heart disease. The anticipated clinical course of CHB secondary to maternal systemic lupus erythematosus is long and persistent, as the conduction system suffers essentially irreversible damage. Furthermore, CHB secondary to structural congenital heart disease is prolonged with the majority of cases needing permanent pacemaking. The major prognostic determinants of CHB are the presence of underlying structural heart disease or congestive heart failure and the ventricular rate. General therapeutic targets for the newborn include supportive treatment for low output congestive heart failure as well as pacemakers for infants with significant bradycardia. Conservative management consists of administration of isoprenaline and accepting a ventricular rate exceeding 55 bpm as hemodynamically sufficient [7].
DISCLOSURE STATEMENTS

Financial Disclosure Statement: All authors have no conflict of interest to declare.

Human Research Statement: Informed consent was obtained for the parents of the twin infants included in the study. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.
REFERENCES


FIGURE LEGENDS

Fig. 1

Chest radiograph taken at two hours of age depicting left-sided tension pneumothorax and pneumopericardium

Fig. 2

Electrocardiograph at 5 hours of age: complete heart block with ventricular rate of 70-78 bpm
Fig. 1
Chest radiograph taken at two hours of age depicting left-sided tension pneumothorax and pneumopericardium

118x94mm (96 x 96 DPI)
Fig. 2
Electrocardiograph at 5 hours of age: complete heart block with ventricular rate of 70-78 bpm

215x110mm (300 x 300 DPI)