Seminars in Fetal & Neonatal Medicine (2005) **10**, 586—593

ELSEVIER

Seminars in FETAL & NEONATAL MEDICINE

www.elsevierhealth.com/journals/siny

Perinatal management, counselling and outcome of fetuses with congenital heart disease

Mats Mellander*

Department of Paediatric Cardiology, The Queen Silvia Children's Hospital, 416 85 Göteborg, Sweden

KEYWORDS
Fetal heart;
Echocardiography;
Counselling;
Pregnancy outcome

Summary Prenatal treatment options for fetal heart disease are still limited but pharmacological treatment of fetal tachyarrhythmias is usually effective. Prenatal catheter interventions are likely to be an option in selected fetal cardiac defects in the future. Delivery should be at a tertiary care centre if the need for immediate neonatal transport is anticipated. When a cardiac problem is diagnosed in a fetus, the parents should be counselled by a paediatric cardiologist specialized in fetal cardiology in close co-operation with the obstetric team. The rate of termination is influenced by gestational age at diagnosis, the severity of the heart defect and the presence of associated malformations. In fetuses with isolated cardiac malformations who are in sinus rhythm with good myocardial function and no or trivial atrioventricular valve regurgitation, the risk of spontaneous intra-uterine death is low. Prenatal echocardiography has the potential to improve postnatal survival in infants with critical heart defects, especially those with duct-dependent systemic or pulmonary circulations.

Perinatal management

Prenatal treatment

The prenatal treatment options for fetal cardiac disease are still limited. They can be subdivided into treatment of fetal tachyarrhythmias, fetal complete heart block and prenatal catheter interventions.

Treatment of fetal tachyarrhythmias

The efficacy of transplacental pharmacological anti-arrhythmic treatment is established in fetal tachyarrhythmias. Although there are no randomized studies, the accumulated experience is convincing. Without treatment, continuous tachycardia usually leads to fetal heart failure with hydrops and fetal death. A prerequisite for effective treatment is correct description of the type of arrhythmia. This can be achieved using either M-mode or Doppler echocardiography. Almost all fetal tachyarrhythmias are supraventricular, and re-entry tachycardia is most common followed by

* Tel.: +46 313434659 / +46 705530606; fax: +46 31845029.
E-mail address: mats.mellander@vgregion.se

1744-165X/$ - see front matter © 2005 Elsevier Ltd. All rights reserved.
doi:10.1016/j.siny.2005.08.002

atrial flutter and ectopic atrial tachycardia. Digoxin given to the mother will usually convert a re-entry tachycardia or flutter,[1] whereas sotalol may be more efficient in ectopic atrial tachycardia.[2] In fetal hydrops, digoxin is less efficient because of poorer transplacental passage; sotalol is often used instead. Direct fetal treatment through cordocentesis is reserved for those unusual cases that do not respond to transplacental treatment. Recently, 100% fetal survival was reported in 29 consecutive cases of fetal tachycardia with one postnatal death.[2] Precise identification of the type of arrhythmia using pulsed Doppler sampling of flow in the fetal superior vena cava and the ascending aorta and consequent tailoring of therapy contributed to these good results.

Treatment in fetal complete heart block

Fetal bradycardia because of isolated complete atrioventricular block is usually the result of damage to the fetal atrioventricular conduction tissue from placental transfer of maternal auto-antibodies. There is significant intra-uterine mortality in this condition.[3] Attempts to reverse an already established block with transplacental steroid treatment have been unsuccessful with few exceptions.[4] Since only a small proportion of pregnancies in auto-antibody-positive women result in fetuses with complete heart block,[5] selection of fetuses for prophylactic steroid treatment (with the objective of preventing the development of heart block) is not possible on the basis of the presence of auto-antibodies alone. Identification of fetuses at higher risk through measurement of atrioventricular conduction times using Doppler is an interesting possibility that merits further investigation.[6] In spite of the irreversibility of complete heart block, treatment with fluorinated steroids has been associated with improved survival to 1 year of age, probably as a result of a lower risk of other immune-mediated complications such as endocardial fibro-elastosis, myocarditis and hepatitis.[7] Whether the possible advantages of such treatment outweigh the side effects[8,9] remains to be determined in large multicentre randomized studies. There is some evidence of a beneficial effect of treatment with beta-agonists in cases with very low heart rates.[10]

Prenatal catheter interventions

It is well known that some heart malformations may progress during pregnancy. In fetal valvar aortic stenosis, left heart structures may grow poorly and the result can be a newborn baby with hypoplastic left heart syndrome.[11–13] Similarly, in fetal pulmonary valve stenosis, the degree of obstruction may increase as pregnancy advances, even to the degree of pulmonary atresia.[14,15] With the rationale of trying to prevent this intrauterine development in order to increase the chances of a biventricular repair, attempts have been made to balloon dilate the aortic[16,17] or pulmonary[18,19] valve in the fetus with varying success. The major problem with this approach, apart from technical challenges and the risk of serious complications, is how to select fetuses for treatment early enough in pregnancy, before significant hypoplasia of the affected ventricle occurs. Earlier in pregnancy, the predictive accuracy of echocardiography with respect to outcome in terms of uni- versus biventricular repair is likely to be lower. Another possible indication for prenatal catheter intervention is a restrictive oval foramen in fetal hypoplastic left heart syndrome, but results of such treatment have not yet been convincing.[20] Nevertheless, prenatal catheter interventions are likely to be an option for selected cases of fetal heart disease in the future.

Timing of delivery

Preterm delivery is almost never indicated in fetal heart disease. Therapy-resistant tachyarrhythmia with signs of increasing heart failure or heart block with heart failure[21] can be exceptions if the advantage of being able to treat the baby directly with anti-arrhythmic drugs, electroversion or pacing are considered to be greater than the disadvantage of preterm delivery.

Mode of delivery

Normal delivery should be the rule in fetal heart disease. In some cases of fetal brady- or tachyarrhythmias, intrapartum fetal heart rate monitoring problems may be an indication for Caesarean section. Also, in the very unusual situation when the fetus may benefit from exact timing of delivery for the optimal planning of immediate postnatal cardiac surgical or catheter interventions, Caesarean section may be indicated. Most critical heart defects are duct-dependent and can be managed with prostaglandin infusions for the days before surgery. Exact timing of delivery is not of any benefit in such cases. In total anomalous pulmonary venous return with severe obstruction to pulmonary venous flow, treatment with prostaglandins is not helpful and open heart surgery within hours of birth may be the only option. Immediate cardiac surgery in such a case may require exact timing of delivery, depending on