

Neonatal and Fetal Refractory Supraventricular Tachycardia Caused by WPW Syndrome: Transplacental, Direct Fetal Treatment, and Postnatal Management

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Received: December 13, 2024; **Published:** January 08, 2024

Abstract

Supraventricular tachycardia is the most common fetal tachyarrhythmia and if persistent often associated with fetal hydrops which can cause intrauterine and neonatal death. The management varies with each center and is typically based on published case series, institutional experience, gestational age at presentation, the presence and degree of fetal compromise, hydrops or other risk factors, maternal condition, and potential maternal risk from both fetal therapy and early delivery (post-natal therapy). The first case was fetal supraventricular tachycardia (SVT) detected at 30 weeks gestational age (GA). Patient was treated by transplacental digoxin therapy for 10 days with no result. We perform direct fetal intramuscular with amiodarone and then digoxin, which resulted in conversion to sinus rhythm sustained only for two days. We decided to perform early delivery and proceed to post-natal treatment. The result of ECG on perinatal period was WPW pattern or preexcitation consists of a short PR interval and prolonged QRS with an initial slurring upstroke ("delta" wave) in the presence of sinus rhythm. WPW syndrome reverted to normal after propranolol and amiodarone combination therapy. On 4 months of age the patient only received amiodarone and the ECG show normal sinus rhythm with normal growth and development.

Keyword: Neonatal; Fetal Supraventricular Tachycardia; WPW Syndrome

Introduction

Supraventricular tachycardia (SVT) is the most frequent fetal tachyarrhythmia [1]. It is characterized by a 1:1 atrioventricular conduction and is the most common cause of sustained fetal tachyarrhythmia, with heart rates > 200 beats per minute (bpm). The most common mechanism underlying fetal SVT is the presence of an accessory electrical pathway between the atriums and the ventricles. The exact location of this pathway cannot be determined by fetal echocardiography [2]. Fetal tachycardia may cause low cardiac output, ascites, pleural and/or pericardial effusions, skin edema, and in some cases can lead to hydrops fetalis, a severe manifestation of fetal congestive heart failure. Hydrops is identified at presentation or develops in about 40 - 50% of fetuses with SVT [3].

Patients with sustained fetal SVT, or patients with non-sustained fetal SVT with evidence of cardiac dysfunction and/or hydrops, can be treated with transplacental therapy, unless there is some unusual contraindication to medicating the mother [3]. Many medications have the potential to break SVT; they differ in their side effects and their ability to cross the placenta. There are no universal standards for drug dosing or the need for loading doses. Drug dosing is empiric and depends on maternal, as well as fetal, factors. To date, there are no trials that show that one drug is more successful than another, so which drug to start with should depend on the conditions of the mother and fetus as well as the preference and experience [2]. Most of the available literature suggests initial treatment with digoxin alone or in combination with a second-line agent, which would typically be flecainide, sotalol, or amiodarone [3].

In setting of severe therapy-resistant arrhythmia, significant myocardial dysfunction, or progressing hydrops, direct fetal therapy may be considered [3]. Several modes of direct fetal treatment have been reported in the literature, including intraumbilical, intraamniotic, intraperitoneal, intramuscular, and intracardiac routes to rapidly achieve high drug concentration in the fetal compartment [4].

Delivery is a valid option for fetuses at term and near term. Postnatal treatment of tachyarrhythmia is usually effective. Some authors advocate that hydrops in a near-term fetus is a clear indication for emergent delivery. Fetal hydrops can cause the rare maternal mirror syndrome. Severe mirror syndrome or severe preeclampsia indicates delivery at any gestational age [5].

Case Report

A 27-year-old woman referred to our hospital at gestational age (GA) 31 weeks due to fetal supraventricular tachycardia. The patient was nulliparous with no remarkable previous illness or medical condition. Patient was already treated with transplacental digoxin therapy (2 x 0.25 mg) for 10 days at previous hospital without any improvement. We perform fetal echocardiography and supraventricular tachycardia with suspected short VA interval and fetal heart rate (FHR) of 220-240 bpm was seen with 1:1 conduction. Tissue Doppler Echocardiography also showed heart rate of 231 bpm with 1:1 conduction. We didn't find pericardial effusion, ascites, or any other major structural abnormality. However, the estimate fetal weight was 2400 gr (percentile > 99%) which correspond to large gestational fetus. Oral glucose tolerance test was high corresponded to gestational diabetes. Screening for thyroid function and infection was normal.

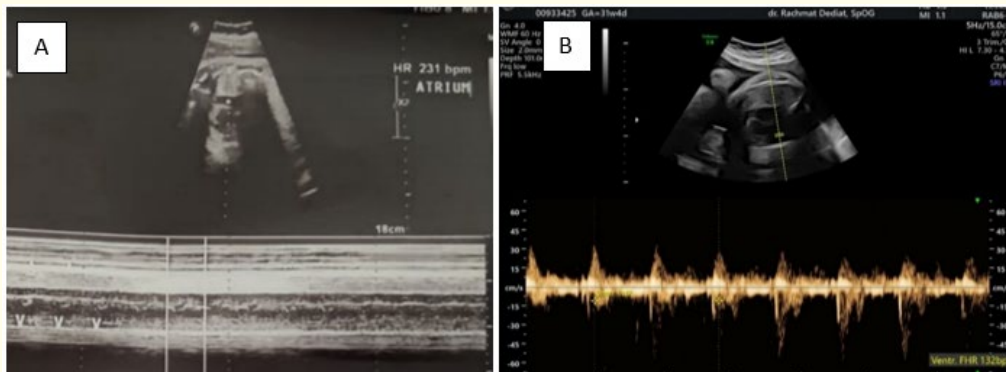


Figure 1: (A) Before treatment: Fetal tachycardia 1:1 atrioventricular conduction. (B) Two hours after treatment: Sinus Rhythm with 132 bpm.

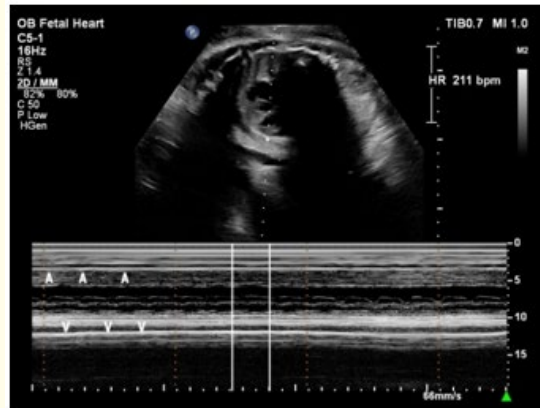


Figure 2: M-mode through RA and LV revealed HR of 211 bpm with 1: 1 on AV association.

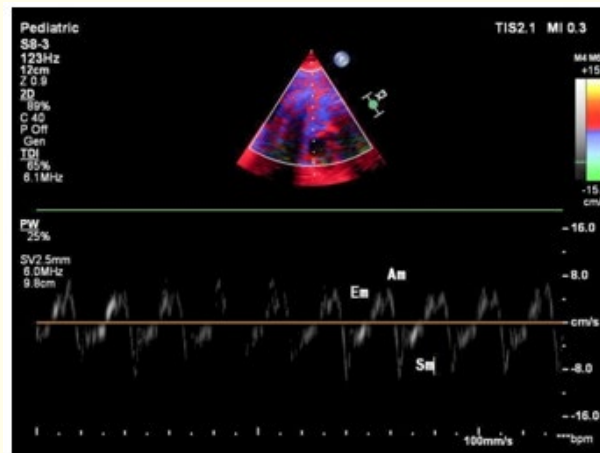


Figure 3: Tissue doppler echocardiography revealed atrial rate (Am) and ventricular A:V rate (Sm) of 211 with 1:1 association.

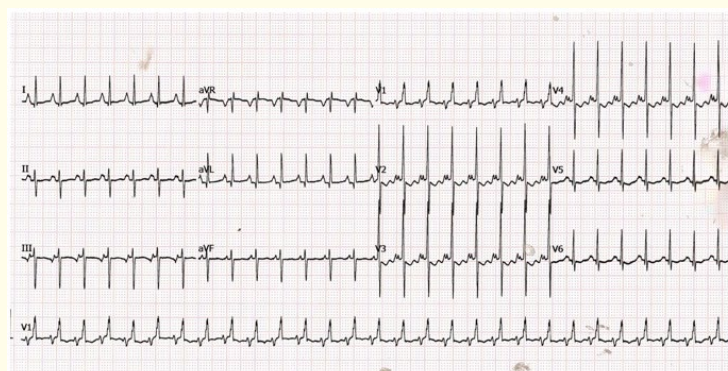


Figure 4: ECG after revert to normal sinus showed HR of 172 bpm, with short PR interval and D wave on V1 which typically of WPW syndrome.

We decided to perform direct fetal antiarrhythmic therapy after counselling and got patient consent. With ultrasound guided needle, we inject 6 mg amiodarone intramuscularly into fetal left thigh. One hour after injection, we achieve cardioversion to sinus rhythm with FHR 132 bpm.

Two days after procedure, FHR was back to > 220 bpm and we decide to perform another direct fetal antiarrhythmic therapy with digoxin. With ultrasound guided needle, we inject 250 mcg digoxin intramuscularly into fetal thigh. After two days of observation, fetal cardioversion was not achieved. We decided to perform early delivery to performed post-natal therapy.

By caesarean section, born 3110 gr, baby Boy with APGAR score 8/8. Post-natal echocardiography and electrocardiography was performed and diagnosed baby with Wolff-Parkinson-White (WPW) syndrome. Baby has persistent and recurrent SVT (HR 200-260 bpm) during hospitalization and treated combination propranolol, amiodarone, and vagal stimulation with cold ice pack. After 4 months of delivery, baby has normal heart rate without delta wave on ECG while still needs amiodarone and propranolol therapy.

Discussion and Conclusion

The management of fetal tachycardias varies with each center and drug therapy is typically based on published case series, institutional experience, the presence of fetal hydrops and gestational age. The optimal outcome is to convert the tachycardia prenatally so that the infant is delivered in sinus rhythm at full-term, without hydrops or other signs of congestive cardiac failure [2].

Patient was referred to out hospital at 31 weeks GA with fetal SVT refractory to transplacental digoxin therapy. We found the FHR was more than 220 bpm which increased risk of developing hydrops. In very preterm fetuses (less than 32 weeks of gestation), combination pharmacological therapy in higher than usual doses or direct fetal treatment with amiodarone, adenosine, or digoxin should be considered instead of delivery if the initial course of transplacental therapy fails [5].

We prefer direct fetal therapy rather than increasing maternal digoxin dose due to several reason. Digoxin has narrow therapeutic window and increasing it doses required maternal toxicity monitoring and serum level concentration which we cannot readily performed [3]. Direct fetal therapy is the recommended management for fetuses at high risk of developing hydrops (sustained tachycardia with ventricular rates more than 220 bpm) [5].

In this case, we successfully achieve cardioversion to sinus rhythm (FHR 120 bpm) 2 hours after injecting 6 mg amiodarone to fetal thigh. We chose amiodarone because it has long elimination half-life and reduces the number of direct fetal injections required to maintain therapeutic drug levels in the fetus [4]. Unfortunately, FHR increased again into more than 220 bpm after 2 days. The next day we perform another direct fetal therapy with 250 mcg digoxin to fetal thigh. We change drug choice to digoxin and hope it can work in synergy with ongoing maternal digoxin therapy. We also found two case report of successful conversion fetal SVT after direct intramuscular digoxin injection [4]. After 2 days of observation cardioversion was not achieved and patient reach 32 weeks GA. After discussion with patient, we decided to perform cesarean section after completing antenatal steroid and begin post-natal treatment for the baby.

After delivery, the baby has refractory SVT (HR 200-260 bpm) during hospitalization and treated the combination of propranolol, amiodarone, and vagal stimulation with a cold ice pack. The neonatal ECG during sinus rhythm showed HR of 172 bpm, with a short PR interval and a D wave in V1 which consistent of WPW syndrome. Digoxin should not be used after establishing diagnosis WPW syndrome, because it can cause AV conduction abnormalities [5]. The latest follow up at 4 months after delivery, baby has normal heart rate without delta wave on ECG. However, babies still need combined amiodarone-propranolol therapy and may need electrophysiological study and ablation of accessory pathways in later life.

Persistent fetal tachyarrhythmias continue to be a challenging entity. The management varies with each center and is typically based on published case series. Direct fetal therapy may be considered if cardioversion does not occur with transplacental treatment, fetal

hydrops or high-risk developing hydrops and fetal gestation is too early to consider delivery. Institutional experience and availability of drug and supporting facilities must be considered.

Wolff Parkinson White Syndrome (WPW) is considered to be a congenital abnormality that involves the presence of abnormal electrical conductive circuits between the atria and ventricles. The disorder includes accessory electrical pathways that bypass the AV node. The hallmark electrocardiographic (ECG) finding of WPW pattern or preexcitation consists of a short PR interval and prolonged QRS with an initial slurring upstroke (“delta” wave) in the presence of sinus rhythm. The term WPW syndrome is reserved for an ECG pattern consistent with the above-described findings along with the coexistence of a tachyarrhythmia and clinical symptoms of tachycardia such as palpitations, episodic lightheadedness, presyncope, syncope, or even cardiac arrest.

WPW pattern arises from the fusion of ventricular preexcitation through the accessory pathway and normal electrical conduction through the AV node. This accessory pathway is thought to arise from chamber myocardium during improper early atrial and ventricular folding in cardiac embryogenesis. As a result, electrically conductive myocardial bundles violate the normal electrical insulation of the atrium and ventricle, forming the accessory pathway. This pathway usually has non-decremental or non-delayed conduction, which is in contrast to the properties of the normal AV node. The electrical conducting characteristics of the accessory pathway can vary and depend upon factors such as the speed of conduction, direction of conduction, and refractory period. These characteristics, along with location and number of pathways, will determine how the pathway may be involved in the initiation or transmission of an arrhythmia leading to WPW syndrome [7]. Asymptomatic patients with WPW pattern, EP study is reasonable, and ablation is reasonable for accessory pathways found to be either at high risk or in patients with high-risk occupations.

Bibliography

1. Çetin C., *et al.* “Successful medical treatment of fetal supraventricular tachycardia that cause hydrops fetalis”. *Turkish Journal of Obstetrics and Gynecology* 11.3 (2014): 193-195.
2. Trisha Vigneswaran and John Simpson. “Fetal arrhythmia’s in: Fetal cardiology: A practical approach to diagnosis and management”. Springer Nature (2018).
3. Malhamé I., *et al.* “Maternal monitoring and safety considerations during antiarrhythmic treatment for fetal supraventricular tachycardia”. *Obstetric Medicine* 12.2 (2019): 66-75.
4. Kang S-L., *et al.* “Foetal supraventricular tachycardia with hydrops fetalis: a role for direct intraperitoneal amiodarone”. *Cardiology in the Young* 25.3 (2015): 447-453.
5. Alina V., *et al.* “Treatment of fetal arrhythmias”. *Journal of Clinical Medicine* 10.11 (2021): 2510.
6. Edgar Jaeggi and Nico A Blom. “Fetal SVT in: Fetal therapy: Scientific basis and critical appraisal of clinical benefits”. Cambridge University Press (2020).
7. Afshar MM., *et al.* “WPW syndrome: Review of the evidence”. *Novel Approaches in Drug Designing and Development* 6.1 (2021): 555679.

Volume 14 Issue 1 January 2025

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