CASE STUDY

Wolff-Parkinson-White Syndrome in Pregnancy, Labour, and Birth: A Case Study

Le syndrome de Wolff, Parkinson et White durant la grossesse, le travail et l'accouchement : étude de cas

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ABSTRACT

Wolff-Parkinson-White syndrome is a cardiac condition which can complicate pregnancy, especially for patients with a new diagnosis. While common management recommendations for labour have been documented, there is very little information about the condition specifically in the case of a new diagnosis, and even less is documented when a patient declines the recommended monitoring during labour. This is a case study of a primip with a prenatal diagnosis of Wolff-Parkinson-White syndrome. She had one episode about two years prior to pregnancy; a second minor episode occurred near the time that she found out she was pregnant, which pregnancy spontaneously resolved and was not investigated; then at 22+5 weeks' gestation, she had a significant episode which spurred testing and diagnosis. Symptoms and common management, including the medical care team's recommendations for pregnancy and labour for individuals with Wolff-Parkinson-White, are discussed. Midwifery care and supporting patient/client-centred informed choice for expectant management versus prophylactic options are explored, including specifically supporting a client choosing care outside of recommendations.This article has been peer reviewed.

KEY WORDS

Wolff-Parkinson-White syndrome, midwifery, patient-centred care, informed choice, case study

This article has been peer reviewed.

RÉSUMÉ

Le syndrome de Wolff, Parkinson et White est un trouble cardiaque qui risque de compliquer la grossesse, en particulier pour les patientes et les patients dont le diagnostic est nouveau. Des recommandations de prise en charge courante ont été documentées pour le travail, mais il y a sur cette maladie très peu de renseignements qui traitent spécifiquement d'un nouveau diagnostic et encore moins de la marche à suivre lorsque la patiente ou le patient refuse le monitorage conseillé durant le travail. Le présent article est une étude de cas d'une primipare avec un diagnostic prénatal de syndrome de Wolff, Parkinson et White. Elle a connu un épisode environ deux ans avant sa grossesse. Un deuxième épisode, mineur, est survenu à peu près au moment où elle a appris qu'elle était enceinte. La grossesse a spontanément résolu le problème, qui n'a pas été examiné. Puis, à 22 semaines et 5 jours de gestation, la patiente a connu un épisode important qui a entraîné la réalisation de tests et l'établissement d'un diagnostic. Les autrices traitent des symptômes et de la prise en charge courante, y compris des recommandations de l'équipe de soins médicaux relatives à la grossesse et au travail des personnes atteintes du syndrome de Woff, Parkinson et White. Elles examinent aussi les soins sage-femme et le soutien du choix éclairé de la patiente ou de la cliente entre la prise en charge non interventionniste et les options prophylactiques, en particulier l'appui d'une cliente choisissant des soins autres que ceux qui sont recommandés.

MOTS-CLÉS

syndrome de Wolff, Parkinson et White, pratique sage-femme, soins axés sur le patient, choix éclairé, étude de cas

Cet article a été évalué par un comité de lecture.

INTRODUCTION

Current literature does not discuss persons with a recent diagnosis of Wolff-Parkinson-White (WPW) syndrome who decline to follow recommendations for the management of labour and delivery.

The client in this case study felt strongly that she did not want to follow all of the recommendations given to her by the medical care team. She had experienced symptoms of her condition only three times; she did not want prophylactic treatment but instead wanted treatment only when and if she experienced symptoms. While supporting our client, we examined the literature for information on the effects of pregnancy and labour on the health of pregnant people and the infant with a (preferably recent) diagnosis of WPW syndrome–specifically, the outcomes of patients who did not follow the recommendations in regard to monitoring during labour. We found no reported cases of death and no cases of morbidity. All the reports outlined recommendations for monitoring that were similar to the recommendations given to our client, and these recommendations were followed in those cases.

We recognize the limitations of a single case study, and the experience and outcome described here is not necessarily applicable to the broader population. However, it is important to share the stories of those who choose not to follow the recommendations but instead to follow a care plan they feel more closely meets their individual needs. Expectant management, rather than a prophylactic approach, is a valued option for those who prefer it. Monitoring and early intervention are often the preferred recommendations for those who are pregnant and have a diagnosis of WPW syndrome. However, the recommended monitoring and interventions can have a negative impact on labour and birth progress and lead to cascading inventions, which in turn can have negative impacts on the child-bearing person and on the infant.

WOLFF-PARKINSON-WHITE SYNDROME IN PREGNANCY

In the early stages of cardiovascular development, normal conduction of an electrical impulse follows a distinct pathway. Typical electrical conduction originates in the sinoatrial (SA) node, located at the junction of the superior vena cava and the right atrium. From here, the action potential spreads through the atria via the interatrial pathway to the atrioventricular (AV) node. The interatrial pathway transmits impulses from the SA node to the left atrium, allowing both atria to depolarize and contract at the same time. The SA node differentiates the atria from the ventricles, so that the contraction and relaxation of the atria occur approximately 0.1 seconds before the contraction of the ventricles. In a heart that is affected by WPW syndrome, a normal conduction pathway is present; however, there is also an accessory pathway. This accessory bundle provides a direct connection between the atria and ventricles by bypassing the AV node and can subsequently cause ventricular pre-excitation.[1]

WPW syndrome is thought to develop during early cardiogenesis when a direct connection between the atria and ventricles is established. During early development, the atria and ventricles have direct contact, although as normal development continues, this contact is broken. If the connection persists, the AV valve defects (termed *muscular bridges*) provide the anatomical base for ventricle pre-excitation, "a condition in which the ventricular myocardium is activated earlier than if the impulse had traveled to the ventricles through the normal atrioventricular conduction system."[2,3] As a result, supraventricular tachycardia and atrial fibrillation are common complications, supraventricular tachycardia being the most common, occurring in approximately 70% of patients with WPW syndrome.[1,3] Atrial fibrillation is said to affect between 15%–32% of people with WPW syndrome.[1,3] In rare circumstances, ventricular fibrillation and sudden death are possible.[3] Most people diagnosed with WPW syndrome also have a congenital heart defect–most commonly Ebstein anomaly–which encompasses a malposition of the tricuspid valve.[2,3] Only 0.1%–3.0% of the general population is diagnosed with WPW syndrome.[1–3] Of those affected, only about half will experience primary symptoms due to arrhythmia.[3] Common symptoms reported by persons with this condition include palpitations, syncopal episodes, shortness of breath, dizziness, and chest discomfort, although most people may be asymptomatic.[3]

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Pregnant women who were previously undiagnosed with WPW syndrome or asymptomatic are at an increased risk of supraventricular tachyarrhythmias when compared with non-pregnant persons, due to the normal physiological changes of pregnancy, such as (1) increased cardiac output, blood volume, and heart rate; (2) increased stress and anxiety acting on the sympathetic nervous system; and (3) increased estrogen, which can alter heart rhythm.[4]

The goal of treating WPW syndrome is to target and block the conduction through the AV node. Treatment ranges f-directed manoeuvres to medication or surgical intervention and depends on a variety of factors, including the type of arrhythmia, how often the arrhythmia is occurring, and the symptoms that are associated with the episodes. Vagal manoeuvres, when practiced appropriately and immediately, can terminate an arrhythmia by slowing the conduction of electrical impulses through the AV node.[3] Vagal manoeuvres that have been effective include the Valsalva manoeuvre (holding the breath while bearing down); an ice-cold wet towel on the face or immersion of the face in cold water; elevation of the lower extremities; elicitation of the gag reflex; and coughing.[3,5] If vagal techniques are ineffective, medications should be used, the first line of which are antiarrhythmic agents, followed by calcium channel antagonists and βadrenergic blocking agents.[1,3] Individuals who are diagnosed with WPW syndrome can be treated when they have episodes or can take an antiarrhythmic agent to reduce the symptoms of an arrhythmia and reduce the possibility of atrial fibrillation. In addition to vasovagal or medical treatment is the option of surgical or transcatheter ablation of accessory pathways.[3]

Of importance, due to the small percentage of persons diagnosed with WPW syndrome in pregnancy, there is not a large amount of data on the risks of syndrome-specific drug therapies. While βblockers used in the first trimester do not increase the odds of congenital anomalies, infants should be monitored for drowsiness, lethargy, changes in sleep, changes in vision, and weight gain after the use of antiarrhythmic. [6,7]

CASE REPORT

The client was a 29-year-old (gravidity, term, preterm, abortion and living [GTPAL] 10000) whose estimated due date was in early 2018. She began antenatal care with midwives at 17+6 weeks' gestation. During her initial visit, she said she planned to have an unmedicated home birth, potentially in water.

Medical History

The client's medical history was significant for suspected WPW syndrome as the client had had one episode of tachycardia approximately two years previously (although no diagnosis was made) and had been asymptomatic since. Her chart indicated that the condition had spontaneously resolved at that time. She had a noncontributory history of an umbilical hernia, which was repaired in 2007 with no complications. She also reported having previously suffered from anxiety, which was treated with medication at that time with no concerns since. At her initial visit with midwives in the summer of 2017, she reported that she had visited a hospital in the spring of 2017 (around the time she found out that she was pregnant) due to a feeling of heaviness in her chest and a racing pulse. She was given a Holter monitor to wear to assess heart function; however, the results were normal, and there were no further tests, follow-up, or concerns at that time.

Obstetrical History

The client's initial laboratory work and ultrasound scans were normal; she declined genetic screening. She complained of ongoing heartburn and indigestion (which were managed with diet) and nausea (which resolved spontaneously without treatment). At approximately 30 weeks' gestation, she was diagnosed with gestational diabetes mellitus, which was well controlled with diet and presented no concern in the pregnancy or during labour.

In September, at 22+5 weeks' gestation age, she had an episode of heaviness in her chest and a racing heart rate while sitting and eating dinner. She attended hospital about 6 hours later upon the recommendation of her midwife, and the on-call obstetrician was consulted. She was assessed in obstetrical triage and found to have a heart rate peaking between 180–220 bpm and ranging from 30–220 bpm with otherwise normal vital signs. An electrocardiogram (ECG) showed premature ventricular contractions and an episode of ventricular tachycardia; WPW syndrome was suspected and confirmed later that same day. A second ECG performed later that day revealed an arrhythmia consistent with WPW syndrome; overall function of the heart was normal. Following obstetrical assessment and observation, cardiology and maternal fetal medicine (MFM) physicians were consulted and had an extensive conversation with the client regarding the risks and benefits to both mother and fetus of reverting the arrhythmia to sinus rhythm with medication. The client agreed to medication, and after she was given procainamide (1 g IV) and metoprolol (25 mg PO bid) for 48 hours for identified atrial fibrillation with ventricular pre-excitation, normal sinus rhythm was achieved.

Additionally, the client was scheduled to have an ablation in two days. She was then admitted to the critical care unit for monitoring owing to elevated troponins (measured in the blood), which can be a sign of damage to the myocardium. Upon discharge two days after admittance, the client was prescribed a β-blocker (bisoprolol 2.5 mg PO qd) for blood pressure control and an antiarrhythmic medication (flecainide 50 mg PO bid). She was referred for a consultation with the London Cardiac Institute Arrhythmia Service to discuss ablation. She was also referred to the pediatric cardiology team for review of the effects on the fetus and the need for fetal monitoring. Follow-up would continue with physicians from the maternal fetal medicine and cardiology departments. A week and a half later, the client was seen in the cardiology department, at which time she disclosed being noncompliant with taking either medication. She explained that she worried about the effects on the baby, and she felt that the medication made her blood pressure drop too much, causing her to feel dizzy. After consultation, she agreed to start taking bisoprolol but was still reluctant to take flecainide. She had a follow-up appointment with a pediatrician specializing in fetal risk assessment from maternal exposure to medications in pregnancy and was reassured of the safety of bisoprolol in pregnancy. However, the client did not feel there was a need for flecainide unless she became symptomatic.

At 25+0 weeks' gestation, the client met with a MFM specialist, and it was determined that her care would be managed by the MFM team. However, the client would continue to be seen by midwives providing supportive care. At 35+1 weeks' gestational age, she was seen in the gestational cardiology clinic, and her β-blocker was switched from bisoprolol to metoprolol, a medication considered safer for breastfeeding. The specialist also recommended cardiac monitoring during labour and oral β-blockers while the client was in hospital for labour. At 38 weeks, the following were strong recommendations from cardiology, the MFM team, and a clinical nurse specialist:

1. Hospital birth
2. Early epidural to reduce the risk of intrapartum arrhythmia from pain and increased catecholamines
3. Continuous electronic fetal monitoring (EFM)
4. Continuous maternal cardiac monitoring done by a critical care unit nurse
5. Induction of labour to ensure the appropriate personnel were present
6. Discussion regarding increased risk that an assisted vaginal delivery could occur during the second stage, to reduce the stress of pushing.

The client agreed to a hospital birth and EFM; however, she declined an early epidural, induction,

and maternal cardiac monitoring. The client expressed on several occasions that she felt that her body would be able to go through labour on its own and that she trusted the birth process. She understood that there were risks with her condition, but because she had not had any further episodes in pregnancy, she felt that it was unnecessary to pursue interventions that seemed, to her, unwarranted. For this reason, she requested that her care be returned to her midwives. She planned to give birth in hospital, where she could labour in the tub and remain upright and mobile throughout her labour as much as possible. She stated she was not opposed to an epidural but wanted to wait until she felt it was an option that she wanted for pain management. The client remained asymptomatic of WPW syndrome for the remainder of the pregnancy. The community standard recommendation is induction at 40+0 weeks' gestational age when a client is diagnosed with diet-controlled gestational diabetes. The client declined this procedure and preferred to let her body enter labour spontaneously. She agreed to undergo increased fetal monitoring with frequent biophysical profiles.

Labour and Delivery

The client was agreeable to an induction at 41+3 weeks—the community standard for all postdate pregnancies—after using natural methods to encourage labour. At 40+5 weeks, a biophysical profile (BPP) done in community gave a score of 6/8 (2/2 for breathing, movement, and amniotic fluid volume, and 0/2 for fetal tone). A repeat BPP two days later was recommended. A repeat BPP at 41+0 weeks resulted in another 6/8 score, this time 0/2 for no fetal movement. After discussions with midwives, the client decided to go ahead with an induction at 41+1 weeks, beginning with cervical ripening with a Foley catheter. The next morning, an amniotomy was performed at 3 cm dilation, and spontaneous contractions commenced. The obstetrical senior resident attended the room and reiterated the identified risk of the client's intention to labour without the recommended cardiac monitoring and epidural analgesia. She encouraged the client to think about the repercussions were she to have an episode of atrial fibrillation in labour—including the risk of mortality for both her and the baby if the attending team was unable to correct the fibrillation. Again, the client expressed her understanding of the situation as well as the risks and declined the recommendations. Four hours later, oxytocin was initiated owing to a lack of progress. A variety of nonpharmacological pain management techniques were used, and the client opted for an epidural at 7 cm. She was found to be fully dilated shortly after the epidural took effect. A vigorous infant was born vaginally after 30 minutes of pushing. The maternal heart rate was assessed every 15 minutes throughout active labour as per her midwives' plan; throughout labour, her heart rate ranged from 90 bpm at its lowest to one episode of 122 bpm while she was drinking coffee. Blood pressure remained normal, and the fetal heart rate ranged from 125–160 bpm throughout the labour. The postpartum period was uneventful.

After her pregnancy in 2018, this client underwent a successful cardiac ablation in the fall of 2019 and is currently back in midwifery care without the additional risk concerns of WPW syndrome. In an update, the client shared that she was very happy with the choices she made regarding her care and that she highly valued being able to trust her body and intuition in making the best choices for herself and her family.

DISCUSSION

The recommendations for this client included many that are commonly accepted for the management of labour and delivery for women with WPW syndrome. This client's goal with regards to her WPW diagnosis and labour was to have few interventions unless she became symptomatic. She wanted to feel heard and supported by her care team. The client was very happy with her care throughout her pregnancy and felt that the positive interprofessional collaboration between team members meant they could provide appropriate care to her and her baby. For some people, the potential risk is worth taking if it means they feel heard and respected in their decisions.

As midwives in Ontario, we speak to the tenet of informed choice—meaning that people may choose to decline recommendations. By providing clients the opportunity to hear the evidence, research, and recommendations from specialists, midwives

can ensure that clients are fully informed and are making a choice that is best for their values and risk tolerances.

SUMMARY

The experience of this client reminds midwives of the importance of focusing care on the client, listening to that client's concerns, and supporting the client's decisions in a proactive and interprofessional manner. Because of the lack of research on WPW syndrome in pregnancy, her team felt inclined to be cautious in the care it provided. However, what proved to be the most effective measure was ensuring that the pillar of informed choice was upheld and respected by all members of the health care team. While this case is unique and while the care and monitoring that were provided are not applicable to all pregnant people with WPW syndrome, providing individualized care rather than following blanket recommendations is valuable and vital in client-centred care.

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