in the post-partum period (up to 12 months), especially for those with LQT2. Beta-blocker therapy was associated with risk reduction in all the studies. Women with LQTS should therefore start beta-blockers at pregnancy or continue beta-blockers at pre-pregnancy dose, with propranolol or nadolol as drugs of choice. Beta-blockers should be continued in the post-partum period, particularly in the case of women with LQT2 due to the increased arrhythmic risk. It should be noted that nadolol has a higher excretion in breast milk than propranolol, with a relative infant dose of 4%–7%.[317] Therefore, nadolol is generally not the preferred beta-blocker during lactation. However, arrhythmic risk can be high in LQTS, and nadolol is one of the drugs of choice; thereofore, a careful weighting of benefit against harm is appropriate. Change of beta-blocker therapy after delivery should be avoided, as this is a vulnerable phase. Therefore, a change from nadolol to propranolol should ideally be evaluated before pregnancy. High dosages of nadolol during lactation may require monitoring of the infant for bradycardia.

Women with LQTS should always avoid QT-prolonging drugs (see www.crediblemeds.org)[318]. Women with LQTS should be promptly treated for hypokalaemia and hypomagnesaemia, which is relevant in pregnancy-related hyperemesis, causing electrolyte disturbances and failure to absorb oral medications. All anti-emetic medications are QT-prolonging. Electrocardiogram monitoring should be performed if anti-emetic therapy is absolutely required.

Long QT syndrome can manifest very early in life, even during the foetal period, and can be a cause of stillbirth[319,320] (*Section 4.5.1*). A neonatal ECG should be performed post-delivery and after 2 weeks to avoid over-diagnosis due to transiently prolonged QT interval during the first 7–10 days of life.[321] Genetic screening for familiar genetic variants should be performed as soon as possible (e.g. from chordal blood). If the newborn is affected by LQTS, beta-blocker therapy should be started immediately.

6.2.2. Brugada syndrome

Men with Brugada syndrome (BrS) are more often symptomatic than women.[322] The only retrospective study on pregnant women with BrS did not show an increased risk of cardiac events during pregnancy and the post-partum period.[323] All patients with BrS should avoid contraindicated drugs (see www.brugadadrugs.org),[324] large meals, or excess alcohol, and promptly treat fever and its causes.[325] If there are symptoms during pregnancy, quinidine therapy should be considered with monitoring of hepatic function and blood count in the mother.

6.2.3. Catecholaminergic polymorphic ventricular tachycardia

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is mainly caused by P/LP in the *RYR2* gene.[252] The only retrospective study published, involving 96 women and 228 pregnancies, did not show an increased risk of cardiac events during pregnancy and the post-partum period.[326]

Beta-blockers are the mainstay of therapy, with additive flecainide if needed.[322,327] Nadolol and propranolol are beta-blockers of choice and should be continued during pregnancy and lactation. As in LQTS, the higher excretion of nadolol in breast milk should be noted (*Section 6.2.1*). Left cardiac sympathetic denervation is a valuable anti-arrhythmic option that should be performed in experienced centres before pregnancy if indicated.[328] Implantable cardioverter defibrillators are indicated in a minority of patients with CPVT.

6.2.4. Short QT syndrome

Short QT syndrome (SQTS) is a rare channelopathy characterized by short QT and increased risk of life-threatening arrhythmias and AF.[329] There are no case reports or studies published on pregnancy in women with SQTS. When choosing an anti-arrhythmic drug during pregnancy, quinidine is the best option in the absence of more robust data.[330]

6.2.5. Labour and delivery in primary arrhythmia syndromes

In all primary arrhythmia syndromes, delivery should be planned with heart rhythm monitoring, electrolyte control, and post-operative ECG monitoring until all anaesthetic drugs have been eliminated.

Labour and delivery may be associated with acute pain, adrenaline release, and urgent administration of anaesthetic drugs, and therefore continuous telemetry monitoring is often warranted. In the absence of obstetric contraindications, vaginal delivery is generally recommended. Neuraxial anaesthesia reduces pain and therefore adrenergic activation which is specifically important in LQTS and CPVT. Women with LQTS and CPVT should continue beta-blocker therapy during labour and delivery. In women with CPVT, it is reasonable to keep the heart rate under the threshold for premature ventricular contraction onset during delivery, typically 100–110 b.p.m.[331,332] Anaesthetic drugs for LQTS should be selected according to the CredibleMeds website (www.crediblemeds.org).

In BrS, propofol and local anaesthetics with sodium-blocking agents (e.g. lidocaine) carry a theoretical risk of triggering arrhythmias. Case reports have indicated uneventful delivery with neuraxial anaesthesia in women with BrS.[333] Thiopental and inhalation anaesthesia have so far not been associated with adverse events.[324,333] Anaesthestic drugs should be chosen according the BrugadaDrugs website (www.brugadadrugs.org).

Recommendation Table 6 — Recommendations for primary arrhythmia syndromes and pregnancy (see Evidence Tables 4–6)

Recommendations	Class[a]	Level[b]
Monitoring and treatment of hypokalaemia and hypomagnesaemia is recommended in pregnant women with primary arrhythmia syndromes suffering from hyperemesis.[334]	I	C
Long QT syndrome		
Beta-blockers[c], with pre-pregnancy dose and with nadolol and propranolol as drugs of choice, are recommended during pregnancy in women with LQTS.[312–316,335]	I	B
It is recommended to continue beta-blocker therapy during lactation in women with LQTS to reduce arrhythmic risk.[148,336]	I	B
Pre-pregnancy beta-blocker dose of nadolol or propranolol, is recommended in women with LQT2, particularly in the post-partum period, which represents a high-risk period for life-threatening arrhythmias.[148,313,315]	I	B

Continued