Association des Malades des Vaisseaux du Foie (AMVF)
Hôpital Beaujon, Service d'Hépatologie
100 Boulevard du Général Leclerc – 92118 CLICHY cedex
www.amvf.asso.fr

PREGNANCY IN PATIENTS WITH VASCULAR LIVER DISEASE

Professor Pierre-François Ceccaldi
- Head of the Gynecology and Obstetrics Department, Hôpital Beaujon, Clichy
- Centre expert des maladies maternelles hépatiques et digestives
- Université de Paris

Doctor Audrey Payancé
- Hospital practitioner in the Hepatology Department, Hôpital Beaujon, Clichy
- CRMVF (centre de référence des maladies vasculaires du foie)

Patients with vascular liver diseases are often women of childbearing age. When a vascular liver disease is known and properly stabilized, the question of pregnancy may arise. Pregnancy is generally a situation that can both promote venous thrombosis and bleeding. These risks may be increased in patients who have already experienced venous thrombosis and who are often treated with anticoagulants. Thus, for these patients, pregnancy must be particularly anticipated and monitored.

Provided that the disease is known, treated, and stable, it is rare today to contraindicate a pregnancy. Indeed, European studies on women with vascular liver disease have shown that the evolution of these pregnancies is good when the disease is known and controlled. In this situation, it is advisable to consult one's hepatologist, gynecologist, and possibly hematologist as soon as there is a desire for a child, before conception. During these pre-conceptional visits, the main elements discussed are the influence of the maternal disease and its treatments on the pregnancy and, conversely, the possibility of a modification of the follow-up of the disease. During these consultations, the specialists ensure that the liver disease is well controlled. An upper gastrointestinal endoscopy should be performed in the year preceding the pregnancy.

The specialists inform the patient about the evolution of these pregnancies. Indeed, it seems that in comparison with the general population, the risk of miscarriage before 20 weeks is increased. In patients followed up and with a stable disease, the prognosis for the mother and the newborn is excellent, although there is an increased risk of prematurity.

Finally, the possible problems of side effects of drugs on the fetus are discussed. Indeed, an adaptation of the anticoagulant treatment is necessary because certain treatments such as antivitamins K can be a source of fetal malformations or fetotoxicity. This is also an opportunity to give a prescription for folates in anticipation of the pregnancy.

Therefore, the pregnancy project must be anticipated and discussed depending on the stability of the maternal disease and the eradication of its complications, such as esophageal varices. The regular follow-up itself is done with a obstetrician-gynecologist and a hepatologist. Childbirth preparation courses by a midwife should also be offered in view of a physiological delivery because, despite the maternal liver disease, it is still favorable to give birth by natural means.

End-of-pregnancy monitoring requires weekly visits to get the best situation (i.e., a cervix that is beginning to open) and temporarily suspend anticoagulant treatment. Treatment with beta-blockers, often prescribed for vascular liver disease, requires monitoring the child's heart rate and blood sugar at birth.

After delivery, follow-up will include monitoring the reintroduction of anticoagulant treatment and monitoring any bleeding (including the return of menstruation). The resumption of oral anticoagulant therapy does not necessarily prevent breastfeeding. It is important to discuss quickly after delivery the type of contraception, either local, by intrauterine device, or by micro-progestogen pill. Finally, an appointment with the hepatologist is usually organized three months after delivery.

Pregnancy in patients with vascular liver disease is possible if it is planned and if the disease is known and stabilized. A follow-up of the pregnancy and the delivery within a specialized and multidisciplinary team is essential for the good progress of these events.

Two patients, members of the AMVF, agreed to share their experiences.

A big thank you to Jean Marie Combes, who collected the testimonies of Sophie and Xiaolei, and to both of them for sharing their experiences with us.

Meeting with Xiaolei and Nicolas, a nice couple. Xiaolei is suffering from portal thrombosis. The life journey to manage her disease and motherhood deserves to be told.

Xiaolei is Chinese and decides to come to France in 2007. Her diploma equivalences obtained at the University of Guangzhou (Canton) allow her to enroll at the Sorbonne to prepare a Master 1 and 2 in tourism. On the cultural level, the pressure is enormous, acquisition of French, way of life, competition with other students, etc. She passes her Master 1 brilliantly. No health problem until July 2009, when violent abdominal pains lead her to go to the emergency room of Port Royal, where painkillers are prescribed without success. The pains persist with more and more violence, and an emergency hospitalization at the Cochin Hospital takes place. After two days of hospitalization, Xiaolei is transferred to Beaujon Hospital, where the diagnosis is made: portal thrombosis. A treatment based on anticoagulants is put in place. Xiaolei is told that this treatment is for life. The stay at Beaujon lasts two weeks, a very difficult passage; loneliness in the hospital, it is necessary to inform the family in China. The Master 2 exam is questioned. Meetings with the psychologist are necessary. The treatment solves a precarious situation. The Master 2 is brilliantly passed.

2013: Nicolas appears in Xiaolei's life. A couple is formed with the desire to start a family. To consecrate this new situation Xiaolei and Nicolas go to China to visit the family. This is the opportunity for Xiaolei to meet Chinese doctors who seem to know little about her pathology and suggest that motherhood will be forbidden to her forever. It is a tragedy; the Chinese culture devalues those who cannot procreate. On her return, Xiaolei goes to Beaujon, where she is told that maternity is possible, provided a rigorous follow-up.

Xiaolei is pregnant. The daily anticoagulants in the form of pills are replaced by injections that Xiaolei manages herself. At the fourth week of gestation, pain appears and leads to a new hospitalization of six weeks at Beaujon. At the same time, a blood incompatibility is discovered between Nicolas and Xiaolei (antibody management), which leads to a weekly brain scan of the fetus in Port Royal to ensure the normal development of the baby. By medical decision, the delivery is induced at 37 weeks; Dorian arrives, it is pure bliss.

And already the couple is thinking of expanding the family. They know the path; it is not an easy one.

Xiaolei is pregnant again. In July 2019, she gives birth to a little Océane at 36 weeks at the Port Royal maternity hospital, whom she has the joy of breastfeeding.

She insists on the fact that the medical communication with the Beaujon Hospital was exemplary.

Meeting with Sophie, 36 years old, university professor, married, mother of a 3-year-old boy Hugo. Since 2008, Sophie is suffering from Budd-Chiari syndrome linked to a myeloproliferative syndrome and a portal cavernoma.

Sophie's story begins in 2006. She is in Berlin for her university studies. Life is going well; she meets her future husband Martin, her health is not a concern... 2008: abdominal pains lead her to seek help. Conventional medication to treat stomach problems is prescribed without success, the pain persists. Sophie then enters a cycle of diagnostic and therapeutic wandering. Finally, the diagnosis becomes clear: the liver vessels are affected. Sophie's situation is critical. Where will she be treated? In Berlin, in France? Sophie's parents, doctors, activate their network of acquaintances. Beaujon with its CRMVF (centre de référence des maladies vasculaires du foie) is identified. There is a medical emergency, and medical repatriation by air is organized. Sophie arrives at Beaujon in critical condition. She is taken care of by the multidisciplinary team of the CRMVF. She spends several days in intensive care before leaving for her room for five weeks. Her memories of this stay at Beaujon alternate between lucidity and moments of absence, certainly due to the morphine administered to her. Sophie remembers having built a shell. She is aware that she has just escaped the worst and that everything remains fragile. Sophie isolates herself from external affects. She is mindful of the commitment of the medical profession. She remembers being visited by a member of the AMVF who had the same pathologies and whose message was clear: "follow your treatment, respect your controls, and live."

The stay at Beaujon is over. Sophie is out of the woods; life goes on with a treatment adapted to her pathologies. The ordeal has strengthened the bonds with her partner, Sophie is getting married. The project of enlarging the family is taking shape. Beaujon is consulted, the message is clear: no maternity, the "mother, baby" risks are too high at this stage.

The couple, still wanting to start a family, starts an adoption procedure. Time passes... until the day Sophie is informed that her case has been reviewed by the CRMVF and that maternity is possible, with clearly identified risks.

During our interview, I sensed an emotion when Sophie recalls this exceptional moment when one goes from an adoption procedure to a possibility of motherhood.

2015, Sophie is pregnant; the pregnancy is uneventful. Twice daily injections, which she does herself, have replaced the treatment with pills, and a monthly ultrasound is performed. Sophie appreciates being monitored like a normal mother-to-be. The delivery is carried out under close supervision, by vaginal delivery, and without an epidural. 2016, Hugo is here, Sophie wishes to breastfeed. A concern remains : is the quality of breast milk independent of the medication administered? The doubt is lifted. Sophie will know the joy of breastfeeding. The adoption procedure is cancelled. Sophie's pathologies being acquired, she has no concerns about Hugo's health.

Sophie recognizes that in this life path, the psychological pressure is enormous and that it is necessary to manage it, that it is also essential to take time to deal with it. That the speed of the diagnosis is critical, that the medical profession must be trusted completely.