BAPEN
Putting patients at the centre of good nutritional care

Conception and Pregnancy in Chronic Intestinal Failure

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Successful conception and pregnancy are possible for an increasing number of women of childbearing age living with chronic intestinal failure (CIF) and receiving parenteral nutrition (PN). There have been case reports and series describing the outcomes of home parenteral nutrition (HPN) dependent women becoming pregnant. Enhanced data capture of both successful and unsuccessful pregnancies within national and international CIF databases will give future information for patients and clinicians.

Key points

Pre-pregnancy (conception) counselling

1. In an ideal scenario, clinicians should have an open discussion with their female CIF patients of childbearing age about pregnancy and potential foetal health. This should aid an informed decision about trying to conceive or preventing pregnancy.
2. The chances of becoming pregnant and carrying a pregnancy to term may be reduced due to the underlying illness, treatment and previous abdominal/pelvic/anal surgery. The use/difficulties of in-vitro fertilisation (IVF) may be discussed.
3. The risks of a pregnancy are dependent on the underlying condition, previous surgery and the current health of the patient (including weight and the presence of inflammation).
4. For all patients planning conception, or as soon as the clinical team is made aware of conception, a detailed review of all current medication must be performed to identify any high-risk medications.
5. Standard pre-conception advice of optimising folic acid, stopping smoking, avoiding alcohol, recreational drugs and certain foods (e.g. with a listeria contamination risk) should be given.
6. All women who are thinking about becoming pregnant should have an intake of folic acid of at least 400 mcg daily. The addition of extra folic acid should be considered if the patient is at a high risk of having a baby with a neural tube defect (NTD).
7. Due to abdominal/pelvic/anal surgery/disease, there should be a discussion about the method of delivery (vaginal or caesarean section).

Management of parenteral nutrition during pregnancy

8. Patients should have 4-weekly clinical reviews during all three trimesters by their CIF multidisciplinary team (MDT) whilst maintaining close dialogue with a high-risk obstetric service who will arrange foetal scans as appropriate, and ensure women have individual birth plans.
9. Any CIF complications are proactively identified and managed as risks may be increased in pregnancy (e.g. catheter-related blood stream infection [CRBSI], central vein thrombosis or intestinal obstruction).
10. At every visit the fluid, macro and micronutrients requirements should be reviewed. Blood is taken for haematological profile, urea and electrolytes, liver function, bone profile, magnesium and C-reactive protein. At 3 monthly intervals, in addition, blood is taken for thyroid function, coagulation screen, vitamin A, D, E, B_{12}, folate, ferritin, selenium, manganese, zinc and copper.
11. Safety data on components of the PN regimen should be assessed. Essential fatty acid (EFA) deficiency increases the risk of foetal neurodevelopmental problems and may be more likely with the use of fish oil emulsions. Olive oil and soya bean emulsions are not associated with this. Monitoring of EFAs should be considered.
12. During pregnancy too much vitamin A is toxic to the foetus and may cause physical deformities. Regular monitoring (minimum 12 weekly) is required. Over-the-counter pregnancy supplements should not be recommended if the patient is receiving intravenous vitamin A.
13. If the mother has a pre-pregnancy body mass index (BMI) <18.5 kg/m², weight gain should be 0.5 kg/week in the second and third trimesters. If BMI is normal pre-pregnancy, a weight gain of 0.4 kg/week should be expected.
14. Most women will require an increase in their PN macronutrients during pregnancy by increasing the energy in the PN infusions or having more nights off PN. Consider a modest increase in PN energy by 200-400 kcals /day from pre-pregnancy amounts in the third trimester.
15. Intestinal failure (IF)/HPN centres should develop links with local and regional high-risk obstetric services, so that there is a clear pathway for women who want obstetric counselling, and for when they become pregnant.
16. If appropriate, the need and timing of a caesarean section should be planned.
17. Breastfeeding may be considered and, if so, the energy provision in the PN will need to be increased and maintained until breastfeeding stops.
18. A parenteral nutrition central vein catheter should not be used for treatments during delivery unless no other venous access can be obtained.

Explanations

1. There are no data to provide a clear evidence base of fertility risk and pre-conception care in CIF. There are, however, established guidelines on the management of conditions (e.g. Crohn's disease in pregnancy which may be extrapolated to the HPN population). Although 54% of respondents in an international survey reported they had experience of managing pregnancy in CIF, 60% of healthcare professionals (HCPs) reported they did not feel it was their role to discuss the topic of pregnancy with their patients with fewer than 10% stating they did it routinely. Thus, patients should be counselled on an individual basis by the CIF clinician and obstetric team regarding their fertility and potential risks of pregnancy. In general, patients of child-bearing age should be supported in their decision about a pregnancy, providing it is felt that successful pregnancy is likely to be safe and feasible for the mother and baby.

2. Active inflammation or malnutrition reduce the chances of conceiving. IVF may be possible if an ovum can be retrieved and implanted into the uterus. If difficult to retrieve an ovum, an egg donor may be considered.

3. Target body weight should be achieved, and all micronutrient abnormalities should be corrected. Clinicians must determine and hence advise about the feasibility and safety of a successful pregnancy (for the mother and baby). Sometimes a patient may be advised not to become pregnant.

4. Not all medicines are safe to take when planning/or during pregnancy, therefore a thorough review, often involving the Trust medicines information team, will allow HCPs and patients to receive accurate information on any potential risks. Medications that are either teratogenic or whose pharmacokinetics may be altered by a pregnancy (e.g. anti-coagulation, anti-epileptic, anti-hypertensive or diabetic medication) may require stopping, changing or needing additional monitoring during the pregnancy.

5. Stopping smoking will reduce exposure to harmful gases (e.g. carbon monoxide, hydrogen cyanide, benzene, formaldehyde, nicotine, phenol, polycyclic aromatic hydrocarbons and tobacco-specific nitrosamines), which will reduce the risk of complications in pregnancy and after birth. Smoking is associated with a risk of stillbirth and premature birth.

   United Kingdom (UK) guidelines recommend that for those planning pregnancy and those who are pregnant, the safest option is to avoid alcohol. Alcohol can increase the risk of miscarriage, and foetal alcohol spectrum disorder which includes poor growth, and learning and behavioural difficulties.

   Dietary advice includes avoiding foods such as soft cheeses and if eating cooked-chill ready meals, they must be well heated. This is to avoid the risk of acquiring a listeria infection which causes few symptoms in a healthy adult but, by easily crossing the placenta, may be fatal to a foetus (miscarriage or stillbirth) or a newborn (pneumonia and/or meningitis).

6. This is in line with the general population: patients should be receiving 400 mcg of folic acid daily prior to conception and for the first 12 weeks of pregnancy to prevent NTD (anencephaly and spina bifida). Cerenvit®/Solivito®/Nutratain® contain 414, 400, and 600 mcg respectively of folic acid. Some patients may need more folic acid (e.g. if a previous pregnancy affected by NTD, are taking epilepsy medications, have coeliac disease, diabetes, sickle-cell anaemia, thalassaemia or have a BMI is over 30 kg/m^2.

7. Delivery may be vaginal or by caesarean section and the risks/benefits should be discussed with the high-risk obstetric team.

8. In addition to the nutrition MDT, patients should be referred to and followed by a 'high-risk' obstetric clinic which may include a 'high risk' foetal medicine team. Access to such a clinic was reported by 80% of respondents in the paper published by Bond et al, while three-quarters of participants felt that a specialist IF-pregnancy service improved the care and outcomes for pregnant patients with CIF. Communication between these teams must be comprehensive.

9. The management of the underlying disease and proactively observing for HPN-related complications is imperative. In a French series, maternal complications unrelated to pregnancy included intestinal obstruction in nine patients, cholestasis in one and CRBSI in five patients, one of whom was subsequently treated with a taurolidine-based lock. The authors also highlighted severe complications in patients with chronic intestinal pseudo-obstruction involving septic episodes, obstruction and uterine rupture in one and obstructive uropathy with renal failure in another, which led them to suggest that this group of patients warranted particularly close follow-up.

10. Monitoring vitamins and trace elements is important. Iron deficiency is very common, and an intravenous iron infusion may be required from the second trimester onwards. Vitamin K deficiency (suggested by prolonged coagulation tests), in addition to causing bleeding, may cause foetal skeletal abnormalities. Although iodine deficiency may be common worldwide screening for it is not commonly performed in the UK.

11. In the French series, all patients received lipid with a mixture of olive oil and soybean-based emulsions, with no patients solely receiving a fish oil-based emulsion. Similarly, in the UK series reporting five pregnancies, all mothers received a combination of lipid (less than 20% Intralipid® or Clinoleic®). Theilla et al. also note that their pregnant patients all received lipid based PN (Intralipid® or Clinoleic®) without significant complications. Thus, close clinical and dietetic overview is required during pregnancy to ensure nutritional requirements are met, particularly since oral intake may diminish in the second and third trimesters.

12. Vitamin A can be harmful to the unborn foetus. Over-exposure can cause a range of malformations including spina bifida, cleft palate, deformities of the eye, ear, limb, heart and kidneys. However, vitamin A is vital to health and development of a healthy foetus. Due to the wide range of oral intake and absorption of patients with CIF, a review of current dietary intake is important, and a decision made for an appropriate route for vitamin A supplementation. The World Health Organization (WHO) recommends that vitamin A supplementation should only be given to pregnant women to prevent night-blindness in areas where vitamin A deficiency is severe (not within the UK). The European food safety agency (ESFA) have set a tolerable upper intake level for Vitamin A as 3000 mcg (10,000 IU). However, due to CIF, where oral intake and absorption varies greatly, provision of vitamin A supplementation may be required depending on dietary intake. Use of Vitlipid® (3300 IU per vial) Nutratain® (3300 IU per vial) or Cernevit® (3500 IU per vial) within HPN needs to be closely monitored to provide adequate but not excessive supplementation.

13. Currently there are no UK guidelines on appropriate weight gain during pregnancy. The United States Institute of Medicine (IOM) can be used as a guide and if the patient has a normal BMI prior to pregnancy weight gain should not exceed more than 0.4 kg/week in the second and third trimesters. Weight gain in a pregnancy in a woman of a normal starting weight is 10-12.5 kg; most weight is put on after week 20. The publication by Billiauws et al. reported median BMI pre-pregnancy of 19.4 kg/m^2, with a median weight gain of 10 kg (IQR: 5) with 33% gaining > 12 kg.

14. In the Billiauws publication parenteral energy needs increased from 5 to 7 nights; 70% of energy needs were met by PN at conception and this increased to 135 % by the end of pregnancy (note median lipid provision/week was 230 g at conception and this increased to a median of 350 g by the end of the pregnancy.
15. Integrated care between obstetrics, foetal medicine and the multidisciplinary NST are vital for providing support.
16. A date for expected delivery needs to be set and if appropriate an elective caesarean section date booked.
17. Breastfeeding is an energy-demanding activity and has to contain enough energy to supply the needs of the growing infant. Therefore, additional energy intake, over and above pre-pregnancy levels, is required (approximately 300-500 additional kcal per day) especially during the first 6 months of lactation.
18. During labour and post-delivery the HPN catheter should not be used for the administration of anything (fluids or medication) except PN. If used for routine or emergency treatment especially by those not familiar with handling these central lines using an aseptic non-touch technique, a CRBSI is likely to occur.

Suggested reading

- Billiauws L, et al. (2017). Pregnancy is possible on long-term home parenteral nutrition in patients with chronic intestinal failure: Results of a long-term retrospective observational study. Clin Nutr.; 36(4): 1165-1169.
- Bond A, et al. (2017). Managing Successful Pregnancies in Patients with Chronic Intestinal Failure on Home Parenteral Nutrition: Experience from a UK National Intestinal Failure Unit. J Gastrointestin Liver Dis.; 26(4): 375-379.
- Theilla M, et al. (2017). Safety of home parenteral nutrition during pregnancy. Clin Nutr.; 36(1): 288-292.
- Bond A, et al. (2023). Experience and opinions relating to pregnancy in patients with chronic intestinal failure: an international survey. Frontline Gastroenterol.; 14(5): 377-383.
- Pironi L, et al. (2023). ESPEN guideline on chronic intestinal failure in adults - Update 2023. Clin Nutr.; 42(10): 1940-2021.
- British Dietetic Association. Healthy Eating During Pregnancy. Accessible online: www.bda.uk.com/resource/pregnancy-diet.html (Jan 2024).
- Food Standards Agency (2022). Lay Summary of the Statement on the effects of excess Vitamin A on maternal health. Accessible online: https://cot.food.gov.uk/Lay%20Summary%20of%20the%20Statement%20on%20the%20effects%20of%20excess%20Vitamin%20A%20on%20maternal%20health#:~:text=Too%20much%20vitamin%20A%20can,heart%2C%20thyroid%20gland%20and%20skeleton (Jan 2024).