Pregnancy and Extracranial Shunts: Case Report and Review of the Literature

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This case involves a 32-year-old pregnant patient with a preexisting ventriculoperitoneal shunt. At term, she had a vaginal delivery with vacuum assistance, an epidural, and prophylactic antibiotics. Although there are no controlled studies validating the best course of management in such cases, there seems to be agreement that vaginal delivery can be attempted in the absence of other obstetrical indications for operative delivery. Obstruction of the shunt and, less commonly, abdominal cysts related to the distal end of the shunt are the principal obstetrical complications associated with a cerebrospinal shunt.

Key words. Pregnancy; pregnancy complications; ventriculoperitoneal shunt.
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There are no controlled studies in the literature about the management of pregnancy in women with ventriculoperitoneal (VP), ventriculoatrial (VA), ventriculo-pleural (VPL) or lumbo-peritoneal (LP) shunts, which are collectively known as extracranial or cerebrospinal fluid (CSF) shunts. However, including the present case, there are case reports of 38 patients involving 51 pregnancies.[1–16] Since it is quite unlikely that there is a sufficient number of cases for a controlled study, management should be based on experience reported in the literature, even though this body of information represents only what is considered to be most safe rather than that which has been proven safe.

As an increasing number of women with CSF shunts are treated for hydrocephalus, those who become pregnant will also require perinatal services. In cases involving maternal hydrocephalus caused by a genetically associated neural tube defect, the fetus should be screened for neural tube defects and hydrocephalus by maternal serum alpha-fetoprotein and serial ultrasonography. When either parent has spina bifida cystica, the risk of the fetus having it is approximately 3%. Other causes of maternal hydrocephalus, such as Dandy Walker deformity, stenosis of the the aqueduct of Sylvius, and cerebellar agenesis, may also have a genetic component.[17,18]

Case Report

The patient in this report is a 32-year-old Hispanic woman, gravida 3, para 4, aborta 2, who was initially seen in the emergency department after collapsing. She was diagnosed as having a tension headache. Eleven days later, she was seen in the physician's office with persistent headache, generalized weakness, and dizziness. Neurologic examination was significant for difficulty in standing up and for unsteadiness in gait and balance. The patient was referred for a head computed tomography (CT) scan, but did not comply with this recommendation. Two weeks later, she came to the emergency department again for severe headaches. Head CT at that time showed moderately dilated ventricles and a large cyst at the third ventricle and lateral ventricle (Figure). The patient's condition was diagnosed as obstructive hydrocephalus secondary to the cyst. A neurosurgeon placed a ventriculoperitoneal shunt with a medium pressure Holter's valve (30 to 45 mm Hg) and performed ventriculostomy of the lateral ventricle with aspiration of the cyst and insertion of a ventricular catheter. The cyst

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Figure. Dilated ventricles with an obstructing cyst secondary to cysticercosis, indicated by the arrow.

fluid had a 1:16 titer for cysticercosis, and the patient was treated with praziquantel for neurocysticercosis.

Sixteen months later, the patient was seen for upper quadrant pain on the right side. She was found to be 10 weeks pregnant, and an abdominal ultrasonogram revealed gallstones. During the course of her pregnancy, she had intermittent right-sided upper quadrant pain, but it was neither persistent nor accompanied by fever. She also reported headaches that were different from those she had previously experienced. In addition, she had had one episode of cystitis, and a weight gain of only 8 lb (prepregnant weight, 150 lb [68 kg]; height, 62 in. [157.5 cm]). The patient was referred for another head CT and follow-up with the neurosurgeon, but again, she did not comply.

At 39 weeks, the patient went into active labor. A follow-up obstetrical consultation concurred with the plan to deliver vaginally with epidural analgesia, to administer 1 g of prophylactic cefazolin every 8 hours for 24 hours, and to not allow the patient to push. She gave birth to a 3220-g female infant with the assistance of a vacuum extractor.

Discussion

Newly diagnosed hydrocephalus is much more common in children than in adults. In children, the most common causes are prenatal or related to malformations within the brain. In adults, the causes are, in descending order, subarachnoid hemorrhage, idiopathic, head injury, tumors, prior operations, aqueduct stenosis, and meningitis. Hydrocephalus may be caused by either increased production of CSF or, more often, obstruction of the flow of CSF. If the obstruction cannot be safely removed, placement of a shunt is a relatively safe alternative. The efficacy of extracranial shunts has improved with the development of new inert materials and better designed valves. Initially, VA shunts were used more commonly. These shunts have unidirectional valves and an implantable flush pump, which can pump CSF through the distal tube when pressed. After problems associated with peritoneal adhesions were solved, VP shunts were used more often. These shunts have the advantage of permitting greater growth, since the length of tubing in the peritoneum is longer than that in the atrium with a VA shunt. This advantage allows children and adults to have a relatively normal life expectancy.[19,20]

The pregnancy of a woman with an extracranial or CSF shunt differs from that of a normal pregnancy, primarily during the third trimester. The areas of concern are shunt functioning, mode of delivery, and risk of shunt infection. Intra-abdominal pressure is thought to be highest in the third trimester because of the enlarging uterus. During labor is the other period when intra-abdominal pressure is thought to be high.

Shunt Complications

Obstruction of the shunt, infection, and problems related to either the proximal or distal end of the tubing [19] are the most common complications associated with CSF shunts. Two reported complications of CSF shunts in pregnancy are obstruction of the shunt and postpartum abdominal complications.

Obstruction may be related to a nonfunctioning shunt or a mechanical obstruction at either end of the shunt. Up to 80% of patients with shunts experience malfunctions, and more than one half of these are related to obstruction.[9] Of the 38 patients with 51 pregnancies, 18 pregnancies were associated with headache, 9 with lethargy, 8 with nausea and vomiting, 3 with ataxia, 3 with gaze paralysis, 4 with new seizures, and 2 with seizures related to subtherapeutic levels of anticonvulsant medications. Since some of these symptoms are also common to both pregnancy and increased intracranial pressure, the patient's history may not provide a clear differentiation between the two. Thirty-eight of 51 pregnancies involved VP shunts, 5 had VA shunts, 6 had LP shunts, and 1 had a VPL shunt. The type of shunt used in the remaining case was not reported.[1–16] In the 4

patients who had surgery for symptoms of persistent shunt obstruction, no actual mechanical obstruction was observed.[4,5,7,10]

It is thought that flow of CSF in pregnant patients decreases either because the shunt is being compressed between the gravid uterus and other internal organs or because intra-abdominal pressure has increased secondary to the large size of the uterus. Eight of the 36 preexisting shunts showed definite signs of malfunction. All but one of these was a VP shunt; the other was a VA shunt. Most cases of shunt malfunction occurred in the third trimester when the uterus is the largest (24 to 37 weeks; average, 30 weeks).

If the intra-abdominal pressure exceeds the pressure in the catheter, shunt dysfunction may occur. Whenever there is a lack of flow, intracranial pressure may build up. The valves are available in different pressures to allow CSF to drain.[1,5,19] Since the distal end of the catheter drains into the right atrium or superior vena cava, where it should not be be affected by the increases in intra-abdominal pressure, VA shunts may be the best option for patients of childbearing age.[5] However, VA shunt malfunction has been reported in one case in which the women was 8 months pregnant.[4] The cause of this obstruction was not stated, but thrombosis and infection are the most common sources of distal obstruction in a VA shunt.[3]

The symptoms of shunt obstruction may include severe headache (possibly bifrontal), nausea, vomiting, lethargy, visual disturbances, and deterioration of consciousness. While headaches are relatively common in pregnancy, shunt obstruction should be considered a potential cause of those that are severe, or those accompanied by neurological signs, or both. The physical signs may include any of the following: ataxia, nystagmus, gaze palsy, confusion, lethargy, decreased memory, decreased venous pulsations, papilledema, and depressed sensorium. There may also be an inability to compress the flushing device of the ventricular shunt catheter.[3–5]

A diagnostic workup could start with a head CT scan, although there is some support for an empiric trial of flushing the catheter or aspirating the reservoir. If the latter is effective, mild or intermittent obstruction is a possibility, and a CT scan and shunt pressure measurement are indicated. If the CT detects changes, some advocate revising the shunt. In two cases in which the patients' CT scans were normal, conservative management with aspiration of the reservoir and pumping proved adequate until the increased CSF pressure resolved with the delivery of the baby.[3,5,7,14] Repeated reservoir aspirations carry a risk of infection.[9] If there is no improvement with pumping or aspiration of the shunt and the fetus is not viable, the VP shunt may be replaced with a VA or VPL shunt in the third trimester to avoid intra-abdominal effects on the device.[9,14]

Another approach is to do a radioisotope or contrast study of shunt patency. The shunt should be tapped to measure intracranial pressure and a culture performed before a decision is made to revise or replace the shunt. If the shunt is patent, treatment with bed rest, fluid restriction, and, in severe cases, steroids or diuretics has been advocated. If a patient's symptoms recur when steroids and diuretics have been discontinued and the fetus's lungs are sufficiently mature, early delivery should be considered. If obstruction is present, revision of the shunt should be considered.[14]

Although CT is the most effective means of evaluating shunt obstruction, one third of patients with obstruction may have normal CTs. When available, a baseline CT during a stable period may assist in the interpretation.[3] Since magnetic resonance imaging involves no radiation, it is a reasonable alternative to CT.[9]

Since many of the procedures to confirm shunt obstruction are invasive, attempts have been made to develop noninvasive modalities to evaluate how the shunt is functioning. These have included magnetic resonance phase imaging, a technique still in the early stages of development, and Doppler ultrasound, which can measure flow while the shunt resevoir is being pumped.[21,22]

Based on neurosurgical experience, other causes of shunt obstruction include obstruction of the ventricular end from choroid plexus, CSF debris, or gliosis. In VP shunts, infection or patient growth may obstruct the distal end, but only rarely. In VA shunts, infection and thrombosis may obstruct the distal end.[3]

Two postpartum abdominal complications, an "extra-abdominal" or abdominal wall cyst and a perforation of the liver capsule, have been reported. The formation of a cyst is thought to be related to increased intra-abdominal pressure during the third trimester or labor causing the shunt to slip out of the abdomen.[6] Thus, a cyst should be included in the differential diagnosis of an abdominal mass in a pregnant women with a VP shunt. Other reported abdominal complications of VP shunts among neurosurgical patients include: CSF pseudocysts, hernias, cerebrospinal ascites or fluid hydrocoele, and perforations of the viscus, abdominal wall, or vagina. All of these complications are quite rare.

Mode of Delivery

During delivery, it is important to avoid bacterial contamination of the peritoneum, especially in the presence of a VP shunt. Historically, avoiding very high intra-abdominal and intracranial pressures has been recom-

mended. Theoretically, either of these conditions might predispose to an ascending infection, retrograde flow or lack of flow. However, since CSF pressure with straining may exceed 700 mm H_2O and intra-abdominal pressure is between 200 and 700 mm H_2O, as measured indirectly through intravesical pressure during the second stage of labor, it seems unlikely that retrograde flow of CSF would occur since there is still a pressure difference. Although there is a one-way valve in the shunt to prevent retrograde flow of CSF, lack of flow may occur.[1,7,23]

Because of the unlikeliness of retrograde shunt flow occurring, recommendations for sufficient analgesia to prevent straining and forceps use to decrease the length of the second stage to avoid prolonged exposure to high intra-abdominal pressures are questionable. Labor pains and straining elevate intracranial pressure, decreasing even further the likelihood of retrograde flow.[14] However, epidural analgesia does carry the theoretic risk of CNS infection, and is especially inadvisable with meningomyelocele or other neural tube defects.[1] With a functioning shunt, herniation is not a risk.

Surgical delivery is another way of avoiding high intra-abdominal pressures. However, a cesarean section itself might also expose the peritoneum, and thus the shunt, to infection unless an extraperitoneal cesarean section was performed. If it is anticipated that a cesarean might be necessary for obstetrical reasons, an extraperitoneal approach theoretically would be used to avoid contaminating the peritoneum.[3] However, not all physicians have experience with this infrequently used and difficult technique. An alternative would be to perform the standard cesarean section, extending the incision so that the distal end of the shunt could be inspected and avoiding obstruction of the catheter by meticulous technique and liberal irrigation. If sepsis or intra-abdominal bleeding occurs, temporary external placement of the distal end should be considered.[7]

In the 38 patients with shunts, 51 of the pregnancies progressed to delivery. Twenty-seven of these had normal vaginal deliveries, 10 had either forceps or vacuum, and 14 had cesarean sections. All had good outcomes. Of the 14 cesarean sections, 3 were for neurologic reasons, 4 were for obstetric indications, 6 were elective, and 1 was performed because of the paucity of literature about management of patients with shunts. All previous authors have recommended operative delivery only for obstetrical reasons and unstable neurologic status, not solely for the presence of a shunt.[7,9,11,14]

In 51 pregnancies, at least 2 patients had no anesthesia, 7 had general anesthesia, 5 had epidural analgesia, 6 had local anesthesia, and 2 had narcotics only. In a neurologically stable patient, there is probably no need for specific anesthesia or mode of delivery other than that used for usual obstetrical indications. However, it may be wise to use anesthesia that does not affect mental status, so that any changes related to increased intracranial pressure can be detected. If there is increased intracranial pressure, epidural analgesia is contraindicated and general anesthesia should be used. Epidural analgesia should be avoided in a patient with a meningomyelocele.[14]

Infection

Prophylactic antibiotics have been recommended because the presence of a foreign body may increase the risk of infection in pregnancy.[1,2] No infectious complications were noted, however, in any of the reported cases, regardless of whether or not the patients were given antibiotics. Some have suggested that antibiotics are necessary only with cesarean section.[3] However, although only 4 of the 14 patients who had cesarean deliveries received antibiotics, there were no infections in either group.

If prophylactic antibiotics are used, they should cover gram-negative bacteria commonly associated with puerperal infections, and *Staphylococcus albus* or *S epidermidis* associated with CSF shunt-related infection.[1,4] For VA shunts, in which the distal end of the shunt is in the superior vena cava or right atrium, prophylaxis for endocarditis should be considered.[24] For prevention of wound infection and sepsis in neurosurgical patients, 1 g of intravenous cefazolin or vancomycin just prior to surgery has been recommended, whereas in patients undergoing cesarean section, 1 g of intravenous cefazolin after cord clamping has been recommended.[25] Most of the antibiotic regimens described have included ampicillin or penicillin with aminoglycoside intrapartum.[2,4,8,15]

Conclusions

In the case report literature for pregnancy and extracranial shunts, extra precautions do not appear to be necessary unless neurological symptoms are present. In operative deliveries, it is important to inspect the distal shunt as well as to avoid obstructing a peritoneal shunt with clots. Increased intracranial pressure is the most common complication of shunts in pregnancy. Headaches associated with nausea, vomiting, and other neurological signs are justification for a workup. Increased intracranial pressure may be managed conservatively using medical measures if there is no shunt obstruction or ventricle enlargement, but obstruction may require neurosurgery. When medical management fails and the fetus is mature, early delivery may be indicated, but only rarely. Although the numbers are small thus far, all deliveries have had excel-

lent outcomes using routine obstetrical care. There does not appear to be any special need for operative or vacuum or forceps-assisted delivery, prophylactic antibiotics, or analgesia other than for the usual obstetrical indications.

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References

1. Monfared AH, Koh KS, Apuzzo MLJ, Collea JV. Obstetric management of pregnant women with extracranial shunts. Can Med Assoc J 1979; 120:562–3.
2. Howard TE, Herrick CN. Pregnancy in patients with ventriculoperitoneal shunts: report of two cases. Am J Obstet Gynecol 1981; 141: 99–101.
3. Kleinman G, Sutherling W, Martinez M, Tabsh K. Malfunction of ventriculoperitoneal shunts during pregnancy. Obstet Gynecol 1983; 61:753–4.
4. Gast MJ, Grubb RL, Strickler RC. Maternal hydrocephalus and pregnancy. Obstet Gynecol 1983; 62(suppl):29.
5. Hankita JK, Suzuki T, Yamamoto Y, Kinuta Y, Nishihara K. Ventriculoperitoneal shunt malfunction during pregnancy. J Neurosurg 1985; 63:459–60.
6. Nugent P, Hoshek S. Large extra-abdominal cyst as a portpartum complication of peritoneal shunt. J Neurosurg 1986; 64:151–2.
7. Samuels P, Driscoll DA, Landon MB, Ludmir J, McKrisky PJ, Mennuti MT, Gabbe SG. Cerebrospinal fluid shunts in pregnancy, report of two cases and review of the literature. Am J Perinatol 1988; 5:22–5.
8. Hassan A, El Moumani AW. Pregnancy and ventriculoperitoneal shunt, report of a case and literature review. Acta Obstet Gynecol Scand 1988; 67:669–70.
9. Cusimano MD, Filomena MM, Gentili F, Sermer M. Ventriculoperitoneal shunt malfunctioning during pregnancy. Neurosurgery 1990; 27: 969–71.
10. Okagaki A, Kanzaki H, Moritake K, Mori T. Case Report: pregnant woman with a ventriculoperitoneal shunt to treat hydrocephalus. Asia Oceania J Obstet Gynaecol 1990; 16:111–3.
11. Frohlich EP, Russell JM, Van Gelderen CJ. Pregnancy complicated by maternal hydrocephalus. S Afr Med J 1986; 70:358–60.
12. Houston CS, Clein LJ. Ventriculoperitoneal shunt malfunction in a pregnant patient with meningomyelocele. Can Med Assoc J 1989; 141:701–2.
13. Yoffe N, Blumenfeld Z. Gestation and spontaneous delivery in hydrocephalus with ventriculoperitoneal shunt. Harefuah 1990; 118:209–10.
14. Wisoff JH, Kratzert KJ, Handwerker SM, Young BK, Epstein F. Pregnancy in patients with cerebrospinal fluid shunts: report of a series and review of the literature. Neurosurgery 1991; 29:827–31.
15. Olatunbosun OA, Akande EO, Adeoye CO. Ventriculoperitoneal shunt and pregnancy. Int J Gynaecol Obstet 1992; 37:271–4.
16. Landwehr JB, Isada NB, Pryde PG, Johnson MP, Evans MI, Canady AI. Maternal neurosurgical shunts and pregnancy outcome. Obstet Gynecol 1994; 83:134–7.
17. Carter CO, Evans K. Children of adult survivors with spina bifida cystica. Lancet 1973; 2:924–6.
18. Habib Z. Genetics and genetic counselling in neonatal hydrocephalus. Obstet Gynecol Surv 1981; 36:529–34.
19. Leech RW, Brumback RA. Hydrocephalus: current clinical concepts. St.Louis: Mosby–Year Book, 1991.
20. Black PM, Ojemann RG. Hydrocephalus in adults. In: Neurological surgery. Philadelphia: WB Saunders, 1990.
21. Drake JM, Martin AJ, Henkleman RM. Determination of cerebrospinal fluid shunt obstruction with magnetic resonance phase imaging. J Neurosurg 1991; 75:535–40.
22. Flitter MA, Buchheit WA, Murtagh F. Ultrasound determination of cerebrospinal fluid shunt patency (technical note). J Neurosurg 1975; 42:728–30.
23. Hopkins EL, Hendricks CH, Cibils LA. Cerebrospinal fluid pressure in labor. Am J Obstet Gynecol 1965; 93:907–16.
24. Prevention of bacterial endocarditis. Med Lett 1989; 31:112.
25. Antimicrobial prophylaxis in surgery. Med Lett 1992; 34:5–8.