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Case Report & Case Series

Management of maternal ventriculo-atrial shunt malfunction during pregnancy

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ABSTRACT

Hydrocephalic females reaching childbearing age is increasing due to treatment advances. It has been suggested that ventriculo-atrial (VA) shunts be preferred over ventriculo-peritoneal (VP) shunts during pregnancy. We present a case with multiple VA shunt malfunctions during two separate pregnancies. We treated the patient with a valveless VA shunt during both and were able to achieve near-term deliveries. During the second pregnancy the patient had an emergent caesarian section due to severe hydrocephalus and stunted fetal growth. Delivering the child also relieved her hydrocephalus. Of unclear reasons the right atrium failed as a distal absorption site during both pregnancies, and we must conclude that VA shunts do not necessarily alleviate problems regarding pressure at the distal end of the shunt system but never the less should be considered a treatment option on a case-by-case basis. Furthermore we conclude that a valveless shunt should be considered in select cases of maternal shunt malfunction where valves exert to high pressure resistance.

1. Introduction

Since the introduction of cerebrospinal fluid (CSF) shunts, hydrocephalic women reaching the childbearing age has become common. The ventriculo-atrial (VA) shunt has been suggested a viable option for patients when ventriculo-peritoneal (VP) shunts fail during pregnancy due to increased intra-abdominal pressure. We present a case with multiple VA shunt failures during two separate pregnancies.

2. Case report

A 31-year old nulliparous female with congenital hydrocephalus was initially treated with a VP shunt. In 2010, after a year with multiple shunt revisions, she developed a shunt infection and peritonitis. The shunt was removed, an external ventricular catheter was implanted, and the infection was subsequently treated with antibiotics. A VP shunt was not considered a viable option due to anticipated abdominal absorption issues secondary to her infection. A differential pressure-regulated VA shunt with an adjustable valve was implanted instead (Strata®).

3. Pregnancy #1

In 2011 the patient became pregnant with in vitro fertilization (IVF). At 24 weeks she developed symptoms of shunt dysfunction and a Computer Tomography (CT) scan confirmed hydrocephalus. A shunt revision was performed that revealed no dysfunction of either ventricular or atrial catheter (an acceptable inflow resistance of 7 cm H_2O to the right atrium was confirmed perioperatively). This led us to conclude the valve was faulty and was therefore exchanged (Strata®). During the following weeks multiple adjustments to the valve where performed to decrease the flow resistance because of persistent headaches and nausea, this however did not relieve the symptoms. A transthoracic echocardiography showed the shunt catheter correctly placed in the right atrium but revealed no pathological findings. Another CT scan was performed that confirmed hydrocephalus. Initially an intracranial pressure (ICP) device was implanted that showed ICP ranging from 8 to 22 mm Hg depending on head positioning whilst on the operating table. Afterwards a shunt revision was performed that, similarly to the previous revision, revealed no dysfunction of either ventricular or atrial catheter. It was concluded that either the valve was dysfunctional or exerted too high pressure resistance even at a low setting. The valve was explanted leaving the patient with a valveless VA shunt. Postoperatively her symptoms subsided and she was discharged. At 38 weeks pregnant she had an elective caesarian section performed due to stunted fetal growth, delivering a healthy child weighing 2700 g. Four months postpartum the patient developed

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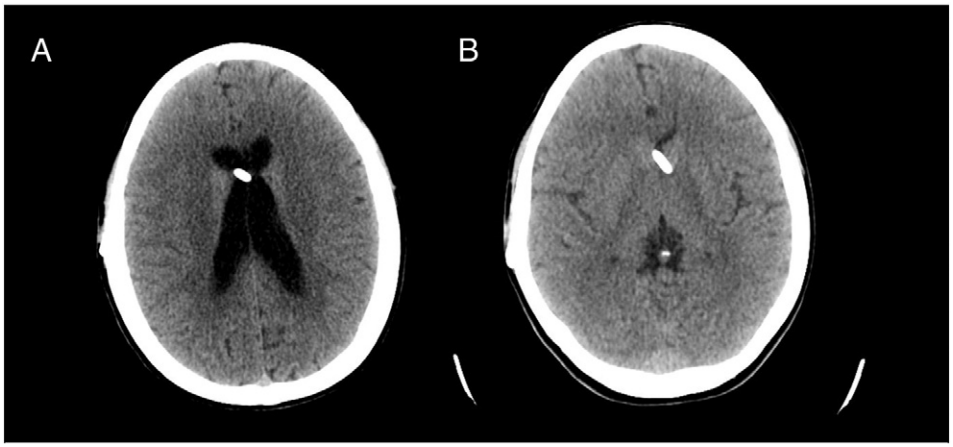

Fig. 1. A) CT scan performed prior to emergency caesarian section showing dilated lateral ventricles and hydrocephalus. B) CT scan 2 months post-partum when patient was asymptomatic.

headaches which intermittently where relieved when positioned supine. Overdrainage was suspected and confirmed by CT scan that showed slit ventricles. A shunt revision was performed where an adjustable valve (Codman Certas®) was implanted into the VA shunt system. Subsequently the shunt was adjusted several times over the course of the next two months in order to find a suitable setting.

4. Pregnancy #2

In late 2013 the patient became pregnant for the second time also with IVF. At 8 weeks pregnant the patient developed headache and nausea and had a CT scan performed that confirmed hydrocephalus. Learning from past experience during her first pregnancy, a shunt revision was performed explanting the valve, and once again leaving her with a valveless VA shunt. This relieved her symptoms. At 28 weeks pregnant the patient was admitted emergently with headache and nausea. She developed drowsiness and a CT scan confirmed hydrocephalus. The shunt was externalized (converted to an external ventricular catheter) for one week, after which a valveless VA shunt was re-implanted. At 38 weeks pregnant her pressure symptoms reemerged and a CT scan confirmed hydrocephalus (Fig. 1A). It was decided to perform an emergency caesarian section and the patient delivered a healthy child albeit with impaired growth (2400 g). Within a day after the procedure her pressure symptoms subsided. At follow up in the out-patient clinic 2 months later the patient was asymptomatic and a CT scan showed no signs of hydrocephalus (Fig. 1B).

5. Discussion

Hydrocephalic women reaching the child-bearing age is increasing due to treatment advances. Shunts are a useful tool in alleviating symptoms of hydrocephalus but are prone to complications such as mechanical obstruction and infection [1]. VP shunts are generally preferred over VA shunts due to being more easily implanted. It has been suggested that VA shunts be a recommended treatment option and even be the treatment of choice for all hydrocephalic women who have the possibility of becoming pregnant [2,3]. Bradley et al. [4] disagrees with the 1990 study by Okagaki, citing the potentially serious complications associated with VA shunts.

The case presented indicates that increased right atrial pressure alone, or in combination with a valve, is what lead to restricted outflow of CSF from the ventricular system through the shunting device. Of course this is merely conjecture as we have found no evidence of increased right atrial pressure (normal transthoracic echocardiography). The patient had no manifest symptoms of pre-eclampsia, which in the minority of cases can cause increased right atrial pressure [5] however both children were born underweight. Had the patient exhibited compression of the inferior vena cava by the gravid uterus (Inferior vena Cava Syndrome) we would have anticipated decreased right atrial pressure through decreased venous return and thus not a reasonable explanation for the patients VA shunt failures.

6. Conclusion

Our case demonstrates that a VA shunt does not necessarily alleviate problems regarding pressure at the distal end of the shunt system (i.e., peritoneum, right atrium). Nevertheless it is our opinion that replacing a VP shunt with a VA shunt during pregnancy should be considered on a case by case basis. We would however not advocate this replacement without their being other abdominal issues besides the pregnancy, unless of course it was evident that increased abdominal pressure caused the malfunction. Furthermore this case has brought to our attention, and underlined the possible benefits of valveless shunts for select cases of maternal CSF shunt dysfunction where shunt valves exert too high pressure resistance.

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