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Open spina bifida: report of malfunction of shunts in adults

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Introduction: About 80% of the patients with open spina bifida develop hydrocephalus requiring shunts in childhood. During follow up in the adult age routine x-ray studies may show disruption of the shunt or a misplaced distal catheter lying outside the heart or the peritoneal cavity. We report our experiences.

Methods: We reviewed the course of 7 (5 males and two females) meanwhile adult patients with open spina bifida shunted in childhood. 6 patients were at first clinically stable, but a routine investigation disclosed catheters of the shunt were either disconnected or too short. In 1 patient the infected shunt had to be removed and the further need for the shunt was unclear.

Results: 1 patient with a disconnected shunt is well and has only been observed for 4 weeks only. 1 patient is well for almost 3 years after his infected shunt had been removed and never been replaced. 1 patient is stable with a peritoneal catheter much too short. 4 other patients, however, needed shunt revision due again because of intracranial hypertension disclosed by close clinical follow up, EEG recording and fundoscopy.

Conclusion: Conservative treatment with close clinical follow up may be an option when disconnection of the shunt or extraperitoneal or extracardial catheter positions are noticed and the patient is stable. A considerable risk for a gradual increase of intracranial hypertension, however, persists. Overdrainage also remains a risk and may require additional treatment (insertion of gravitational components or adjustable valve systems). A close follow-up and early shunt revisions in this patient group are recommended.