Introduction

Hydrocephalus is a crippling disease of childhood. It results from excessive accumulation of CSF in the ventricular systems of the brain due to overproduction or under-absorption.\(^1\) It is of two types; obstructive and communicating. Obstructive hydrocephalus results from obstruction to the pathways of CSF circulation either within the brain or at the level of basal cisterns whereas communicating hydrocephalus results from failure of absorption of CSF into the venous circulation at the level arachnoid granulations and villi. Hydrocephalus could be congenital due to aqueductal stenosis, Chiari malformations, Dandy-Walker’s malformations, intrauterine viral infections or acquired secondary to childhood tumours like colloid cysts, tectal tumours, pineal body tumours, meningitis (36%), encephalitis, otitis media, dural sinus thrombosis.\(^2-7\) Clinical presentation varies according to the age of onset of hydrocephalus. In infants, it presents with gradually increasing diameter of skull, bulging of anterior fontanelle, diastasis of skull sutures, vomiting, lethargy, feeding problems, sun-setting sign with forced downwards gaze and Macewen’s crack-pot sign on head percussion. In children and teenagers, it presents with chronic early morning headache, vomiting, lethargy, learning problems at school, behaviour problems and intellectual impairment.

Ventriculoperitoneal shunt is the gold-standard for extracranial drainage of CSF at all age groups. Ventriculoatrial and ventriculopleural shunt are used as backup shunts when peritoneal cavity is unaccommodating. Ventriculopleural shunt is rarely used because of its associated risks of respiratory failure, pneumothorax, hydrothorax, empyema and fibrothorax. We report the case of a 12-year old boy who presented to PAEC hospital, Islamabad with complications following insertion of a ventriculopleural shunt for communicating hydrocephalus which developed after surgery and radiotherapy for medulloblastoma.

Case Report

A 12-year old boy, student of 6th class, resident of Rawalpindi, developed headache and vomiting. His CT brain revealed medulloblastoma for which he was operated upon in March, 2004 followed by craniospinal radiotherapy for 5 weeks. Three months after surgery and radiotherapy, he again developed headache and repeated vomiting. His CT-brain was repeated which revealed a communicating hydrocephalus MRI brain revealed no tumour recurrence. He was again operated upon and ventriculopleural shunt was placed in December, 2004. Headache and vomiting disappeared after shunting. He remained reasonably well till June 2006 when he developed breathlessness on exertion which progressed to orthopnoea and paroxysmal nocturnal dyspnoea over 3 week duration. He complained of inability to sleep and loss of appetite of the same duration. His parents noticed episodes of irritability and aggressive behaviour. On examination, he was conscious and oriented. His heart rate was 130 bpm and respiratory rate of 73 /min. He was afebrile with blood pressure of 100/60 mmHg and Glasgow Coma Scale (GCS) of 15/15. He was found in respiratory distress with working of accessory muscles of respiration and of alae nasi. His neck veins were engorged. He was pale but showed no evidence of cyanosis, clubbing, koilonychia or lymphadenopathy. Trachea was shifted to the left and apex beat was palpable in the 6th intercostal space in the anterior axillary line (mediastinal shift). Chest was asymmetrical chest with diffuse bulge on
the right side. Chest movements and expansion were significantly decreased on right side. Percussion note was stony dull on the same side. Vocal fremitus and vocal resonance were markedly reduced on the right hemithorax. Air entry was almost absent on the whole right side both anteriorly and posteriorly. He had a fixed, dilated left pupil with mild ptosis due to palsy of left oculomotor nerve after surgery for brain tumour. Abdomen was soft and non-tender but liver was enlarged 4 cm below costal margin.

Investigations done were CBC, LFTs, RFTs and serum electrolytes which revealed no abnormality. Chest X-rays showed opaque right hemithorax with shifting of mediastinum towards the left side. Distal end of the ventriculopleural shunt was visible in the middle of right pleural cavity. Pleural Tap (Thoracentesis) showed yellow crystal clear aspirate, Protein; 1.8 gm% (transudate), TLC = 70 / cmm, Polys = 5%, Lympho = 95%, Gram and ZN staining were negative for micro-organisms and cytology was negative for malignant cells. The patient was diagnosed as a case of right-sided tension hydrothorax with leftwards mediastinal shift secondary to right ventriculopleural shunt. He was operated upon. His right ventriculopleural shunt was converted to right ventriculoatrial shunt and right-sided chest intubation was done with slow decompression of tension hydrothorax. Postoperative recovery was smooth and speedy with no complications. Air entry improved on the right side and dyspnoea gradually settled. Chest tube was removed on 5th postoperative day when serial chest X-rays showed full-expansion of the right lung and return of mediastinum to its normal original position and tip of distal catheter was visible in right atrium of the heart (Figs. 1-4). He was discharged on 6th postoperative day. His follow-up visits were unremarkable and repeat chest X-rays showed fully-expanded right lung with no pleural effusion.

**Discussion**

There are many ways to treat hydrocephalus like daily lumbar punctures, ventricular taps, pharmacotherapy to reduce the production of CSF by blocking carbonic anhydrase (acetazolamide 100 mg/kg/day) and ventricular shunting. Shunts have become well-accepted standard treatment of childhood hydrocephalus. The development of valve-regulated shunt systems provides a major advance in the treatment of hydrocephalus. The key feature of all shunt systems is that the drainage of CSF from ventricles to a distant site is controlled by a valve-membrane to prevent overdrainage of CSF and development of slit-ventricle syndrome. “Once shunt for ever shunt” is probably true for most of the children. The goal of shunting is to normalize the intracranial pressure and to allow re-expansion of the brain tissue to constitute a cortical mantle that is at least 3.5 cm thick to maximize the child’s development. Shunts prevent progressive brain damage and halt intellectual deterioration and prolong the survival rate in hydrocephalic children. Ten year survival rate is 95% in shunted children as compared to 46% in non-shunted children. Similarly, intellectual impairment is 30% in shunted children as compared to 62% in non-shunted children. Shunting is one of the basic neurosurgical operations with high complication, failure and revision rates. Historically, many sites have been used to accommodate the distal end of the shunts like peritoneal cavity, pleural cavity, right atrium, ureter, gallbladder or fallopi an tubes with pros and cons of each site.

Ventriculoperitoneal shunt has become the gold-standard and preferred choice for extracranial diversion of CSF, especially in infantile hydrocephalus. Ventriculoatrial and ventriculo-pleural shunts are possible alternatives to ventriculoperitoneal shunts. Ventriculoperitoneal shunt is preferred because peritoneal cavity has large absorptive capacity and can retain a good length of drainage catheter (up to 36 inches) allowing for growth potential of the child and less need for shunt revision as the child gains height. Moreover, rhythmic contractions of the abdominal viscera prevent entrapment of distal catheter in the scar tissue or peritoneal adhesions and causing its blockade. However, there are certain complications associated with ventriculoperitoneal shunts like intestinal perforation, peritonitis, intra-abdominal adhesions, intestinal obstruction, ascites, CSF-filled pseudocyst formation. Disconnection, migration and expulsion through umbilicus, vagina or anus have also been reported. Some complications can also occur at proximal end of the shunt catheter like cerebral and ventricular haemorrhage, overdrainage (slit-ventricle syndrome), underdrainage, cognitive decline.

Ventriculoatrial shunts are the second choice in children more than 12 years old. The distal end of the catheter is placed in the right atrium through internal jugular vein. However, there are fatal complications associated with the VA shunt like septicemia (2-8%), infective endocarditis, cardiac arrhythmia, pericardial effusion, cardiac tamponade,
disconnection and migration of distal end into the cardiac chambers. Shunt nephritis with haematuria and proteinuria is also a complication unique to

Fig. 1: Proximal End of Ventriculopleural Shunt
Fig. 2: Communicating Hydrocephalus

of the heart has largely been abandoned as initial choice but it remains as a viable second option when infection or surgery renders the peritoneal cavity unaccommodating for distal catheter.

Ventriculopleural shunts are used as second backup for ventriculoperitoneal shunts in children younger than 12 years of age whenever there are relative or absolute contraindications to creation of ventriculoperitoneal shunt. It was first introduced by Ransohoff in 1954. However, ventriculopleural shunt has failed to gain popularity among neurosurgeons and is seldom used in the management of childhood hydrocephalus because of certain serious complications associated with it like respiratory insufficiency, pleural effusion and pneumothorax. Tension hydrothorax is one of its fatal complications. Actually, the pleural cavity is not meant for absorption. Therefore, gradual accumulation of CSF leads to the development of massive tension hydrothorax with compression of ipsilateral lung and shift of the mediastinum towards opposite side (mediastinal shift) leading to the onset of severe respiratory insufficiency as was the case with our patient. There are also high chances of hydrothorax becoming infected leading to development of empyema thoracis with life long misery. Shunt infection is the most common and dangerous complication with the reported incidence ranging between 2.6% to 38%. The morbidity of shunt infection is severe, a single episode lowering the IQ by 10-30 points. Another serious complication of ventriculopleural shunt is the development of fibrothorax because of gradual sedimentation of CSF-proteins and fibrinous exudates over the pleural surfaces. Sometimes, pleural end of Ventriculopleural shunt gets retracted to drain in the subcutaneous tissue of breast leading to breast enlargement and CSF galactorrhoea. Rarely the tip of the distal catheter causes diaphragmatic irritation and hiccough. Because of these lethal complications, ventriculopleural shunt is least commonly used in the management of hydrocephalus. Sometimes, certain brain tumours like glioblastoma can metastasize to pleural cavity through ventriculopleural shunt and can lead to malignant pleural effusion. Fortunately our patient had no tumour recurrence and no spread to extra-cranial tissue.

Hence, we conclude that the use of ventriculopleural shunt should be avoided whenever possible for CSF diversion as pleural cavity is not meant for fluid absorption and may lead to gradual CSF accumulation and development of massive tension hydrothorax as our case did. However, with the advent of new developments in the valve technologies, ventriculopleural shunts may be used as a valuable alternative for CSF diversion when other sites like peritoneal cavity or heart are unsuitable or unavailable for shunting. The incorporation of an anti-siphon device in the shunting system seems to decrease the possibility of pleural effusion. Such types of devices usually continue to function for over 5 years. Wherever possible the ventriculoperitoneal shunt should be used as the first choice for treatment of childhood hydrocephalus due to its lower incidence
of revision, serious infection and morbidity. Since the long-term complications and mortality of ventriculoatrial and ventriculopleural shunts are higher, these should be used only when there are relative or absolute contraindications for the use of ventriculoperitoneal shunt. Recently, endoscopic Third-Ventriculostomy is fast gaining acceptance as an alternative to CSF shunts with lower complication rate, in the management of obstructive hydrocephalus.

References