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News and Perspectiveson Treatment of NormalPressure InternalHydrocephalus

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Abstract: *Many patients, usually over 60 years old, presenting presenile dementia associated with marked gait disorders, impaired balance, urinary incontinence, have been shown to have enlarged ventricles associated with relatively small cortical atrophy. Intracranial pressure monitoring indicates normal values, or subject to only minor peaks, usually at night. Because some of these patients improve markedly after ventricular shunting procedures it has been suggested that their neurological dysfunction may be caused by a pressure effect on the brain from the increased internal surface of the ventricles. Many of these patients do benefit from surgery, and a lot of them have a history of subarachnoid hemorrhage, traumatic brain injury or meningitis which might have impaired the CSF absorption.*

INTRODUCTION

We would like to present the experienceof our clinic over the last fiveyears regarding the treatment ofnormal pressure internal hydrocephalus(28 patients operated between January2009 and December 2013), to report our results andcompare them with the statistics and results fromthe international literature. Normal pressure internalhydrocephalus (NPIH) represents an increase ofCSF volume, with different etiology, that causes anenlargement of the ventricular system as a consequenceof the hydrodynamic CSF circulation disorders.

The cause of this disease cannot be identifiedin 60% of cases. It was described in 1965 with theHakim & Adams triad: gait disorders and impairedbalance, cognitive disorders (progressive dementia),sphincter disorders (8). The imagistic explorations(CT scan) indicate the size of hydrocephalus; ICP< 15 mmHg and the pressure gradient between theventricles and subarachnoid space is very low. Fortreatment there are extrathecal shunts (particularlyventriculo-peritoneal shunt) and intrathecal shunts(particularly endoscopic ventriculocisternostomy).

Both methods have been practiced successfully inour clinic.

The ideal treatment method for hydrocephalusstill does not exist. None of the surgical techniquesis perfect and no shunting device gave total satisfaction(4, 7).

Hydrocephalus represents the enlargement ofone or more parts of the CSF containing anatomicstructures. The CSF total volume is about 150 ml inadult; the ventricular system contains 25-30 ml, thespinal subarachnoid space contains 30 ml and therest of CSF

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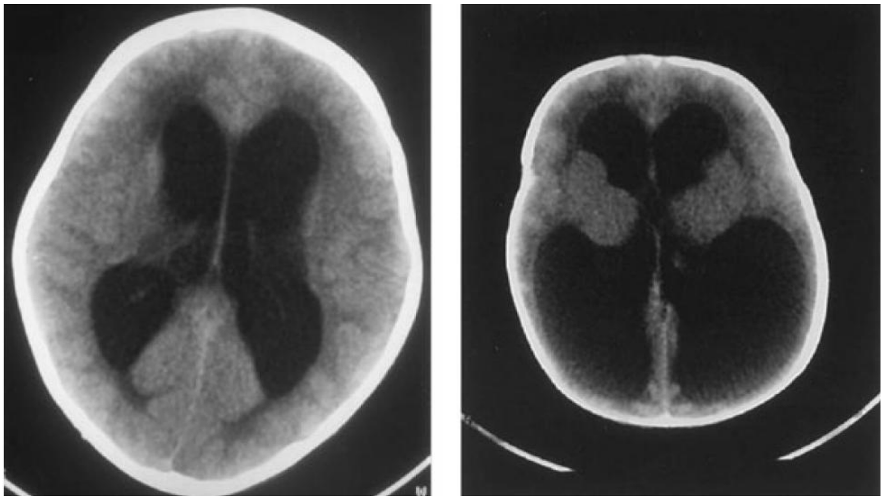
is contained by the cranial subarachnoidspace and basal cisterns (1).

Figure 1. The ventricular enlargement shown on CT scan

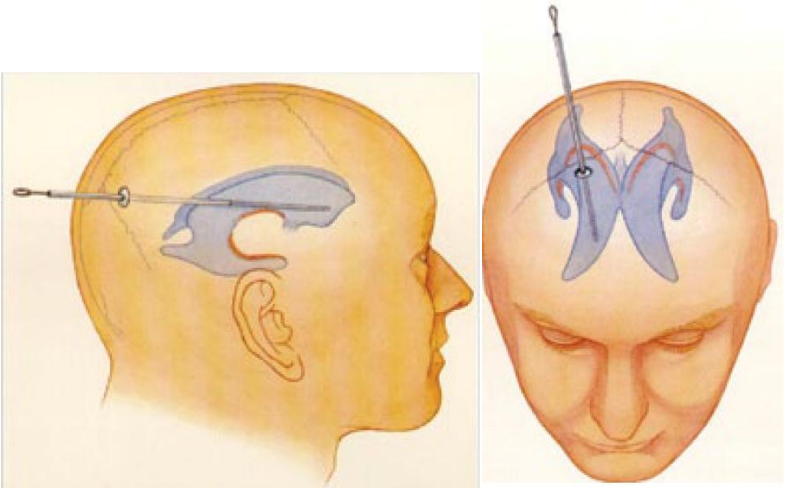


The total production ofCSF is about 600 – 700 ml/24 hours, approximately0,35 ml/min. The CSF participates in maintainingthe endocranial volume constant by adjusting theproduction/absorption ratio depending on the cerebral parenchyma volume and the intracranial bloodvolume variations according to the Monro-Kellierelationship. Changing the volume of one of thethree intracranial components (brain tissue, blood,CSF) is followed by a compensatory reaction fromthe other two components in order to maintain insidean inextensible space the endocranial volumeconstant (9).

Based on its underlying mechanisms, hydrocephaluscan be classified into obstructive and communicating.

The obstructive hydrocephalus is definedby any condition that restricts the CSF flowto and from the ventricular system. Any CSF flowinterruption outside the ventricular system definesthe communicating hydrocephalus (5).

Figure 2. Lateral ventricles puncture



The NPIH etiology is not fully known. Thereare many possible congenital or acquired causes, butthe most important are the subarachnoid hemorrhages(20%), meningitis (1%), parasite infections,traumatic brain injuries, neurosurgical procedureswith open ventricular system, intoxications, Alzheimerdisease (15%).

Hydrocephalus of the adult patients is a communicating,chronic and normal pressure hydrocephalus(3).

DIAGNOSTIC

The CT scan and the MRI revolutionized thediagnosis and the postoperative follow-up of thehydrocephalus. The CT scan is the first stage of diagnosis.

It highlights an obstructive cause, evaluatesthe ventricular enlargement, appreciates the cerebralparenchyma condition (periventricular hypodensity)and the subarachnoid spaces condition (basal cisterns,sylvian fissures, interhaemispheric fissure, corticalsulci).

Bifrontal index measuring (the distancebetween frontal horns / intracranial distance ratio, onthe same CT slice) > 50% is suggestive for a possibledecompensation of hydrocephalus.

The Evans index> 30% has the same meaning (the frontal horns size/ the maximum biparietal diameter ratio). It suggestsan active hydrocephalus. The MRI completes the CTscan by describing the obstructive lesions accuratelyand obtaining dynamic information over the CSFflow (the absence of CSF flow through Sylvius aqueduct).

The preventing treatment is important becauseof the existing risk of postoperative hydrocephalus afterany neurosurgical procedure. Cisternograms withradioactive markers (99Tc-DTPA) remain controversialand rarely used.

The radionuclide is injectedinto the subarachnoid space by a lumbar punctureand serial images are taken by planar scintigraphy 3,6 and 24 hours after the injection. In case of NPIHintraventricular radioactivity can be obtained even48 hours after the injection. Serial lumbar punctureswith repeated evacuation of 15 – 30 ml CSF associatedwith clinical improvement can predict a favorableresponse to shunting procedures. Patients withinitial measured CSF pressure >15 mmHg respondedfavorably after ventriculo-peritoneal shunting (6, 8, 9).

TREATMENT

The treatment of hydrocephalus depends onthe moment of diagnosis, etiology, age and clinicalcondition of the patient (particularly the acuteform) and the complementary investigations results.

The treatment with acetazolamide, a carbonic anhydraseinhibitor has favorable effects predominantlyby inhibiting the chorioid plexus secretion and lessby the diuretic effect.

The acetazolamide dose is 25mg/kg/day with simultaneously administration offurosemide 1 mg/kg/day.

The treatment of obstructive hydrocephalus isremoving the obstacle (excision of tumors). The surgicaltreatment seeks not returning to normal size ofventricles but regaining most of the lost neurological functions.

The diuretic and corticosteroid therapiescomplete the CSF evacuation by lumbar punctions.The CSF lumbar drainage will be performed onlyafter the confirmation of communicating chronichydrocephalus by imagistic methods.

There are several types of extrathecal derivationsof CSF: controlled external ventricular drainage,ventriculoperitoneal drainage, ventriculoatrialdrainage, ventriculopleural drainage.

The currentlyused valves are predetermined opening pressurevalves (low, medium and high pressure), modularopening valves and programmable valves with variableresistances, self-regulating valves etc.

The intrathecal derivation (particularly theendoscopic ventricular cysternostomy) representsan alternative treatment method. Both extra- andintrathecal derivations have been successfully performedin our clinic.

The intrathecal CSF derivations consist of endoscopicventriculostomy through the 3rd ventriclefloor aiming to restore the communication betweenintraventricular and subarachnoid liquidian compartments.

MATERIAL AND METHODS

We retrospectively studied 28 patients admittedto our clinic between January 2009 and December 2013. The patients were all neurologicaland imagistic diagnosed with normal pressure internalhydrocephalus.

Only in 18 cases, a cause forimpaired CSF absorption and ventricular enlargementcould be detected. 8 patients had a historyof traumatic brain injury, 4 patients had a historyof subarachnoid hemorrhage (First and secondgrade on Hunt and Hess scale) with normal "4vessels" cerebral angiogram, 2 patients were diagnosedwith Alzheimer disease prior to admissionand 4 patients had a history of ischemic stroke.

3 of the patients with NPIH after subarachnoidhemorrhage underwent endoscopic procedureswith intrathecal derivations (ventriculocisternostomy).

On the other 15 patients were performedventriculo-peritoneal shunts using various valves(most of them were low pressure valves). The mostfrequently used were the Delta (Medtronic), Spitz – Holter, Heyer – Schulte, Pudenz, Cordis – Hakimvalves. In 10 cases, a cause for NPIH was not revealedand these patients were diagnosed withidiopathic NPIH. They have also been performedventriculoperitoneal shunts with low pressurevalves.

Clinical improvement was significant inmost patients, but only partial in the patients diagnosedwith Alzheimer disease.

According to statistics from literature, urinaryincontinence is the main symptom that resolves aftershunting procedures.

Gait disorders and impairedbalance are subsequently remitted and dementia isthe last that improves (3, 4, 8).

Black and collaboratorsestablished few criteria that can predict thefavorable clinical course after shunting procedures:

- clinical: the presence of symptomatic triad;approximately 77% of the patients presenting gaitdisorders as primary symptom improve their locomotorfunction after shunting; the patients withdementia without gait disorders rarely improve afterdrainage;
- patients with CSF pressure >18 mmHgon lumbar puncture or continuous monitoring improvetheir neurological status after ventriculoperitonealdrainage;
- patients with CT or MRI showing largeventricles with minimal cortical atrophy have favorableevolution after shunting procedures.

The response to drainage is especially good asthe symptoms started recently (1, 5).

ILLUSTRATIVE CASES

Case 1.

66 year old male patient complaining ofgait disorders, impaired balance, cognitive impairment(occasionally) and sphincter disturbances(imperious need to urinate) which started abouta year ago, slowly progressive despite of conservativetreatment. We decided to install ventriculoperitonealdrainage with self-regulating valve. Postoperativeevolution was favorable, symptoms therebyimproving considerably about 3 months after theprocedure.

Case 2.

59 year old male patient complaining of headache, nocturnal insomnia, depressive syndrome,cognitive impairment, gait disorders and locomotor's instability.

Case 3.

68 year old female patient hospitalized for memory disorders, sphincter disturbances (imperiousneed to urinate), gait disorders, vertigo, occasional headaches and depressive syndrome. The CT scan

revealsenlarged ventricular system. It was decided to install VP drainage with Delta low pressure valve. The symptomsimproved partially after one month with significant improvement 6 months after the procedure.

Figure 3. The ventricular catheter placed inside the right lateral ventricle and the selfregulating valve placed in the right parietal region, under the scalp.

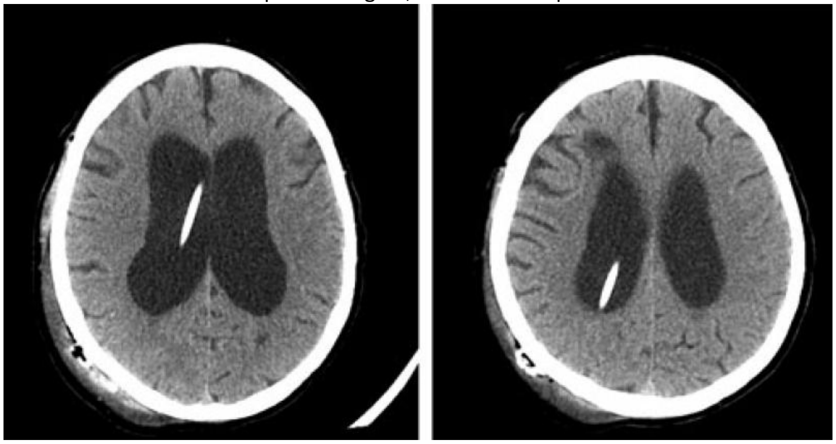
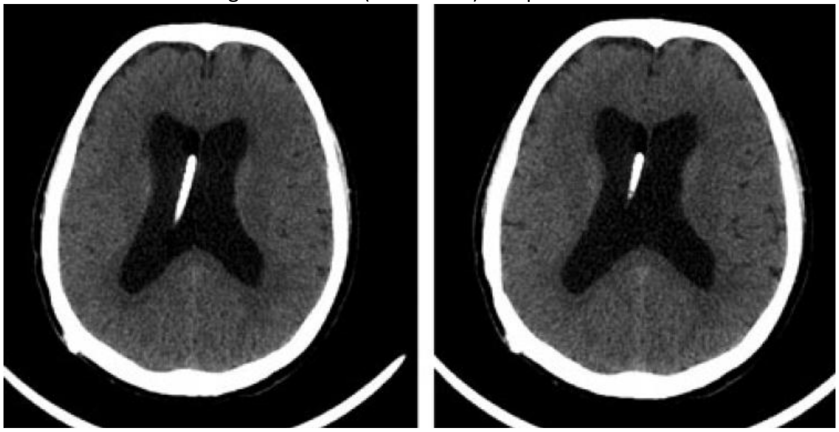


Figure 4. The ventricular catheter placed inside the frontal horn of the right lateral ventricle, near septum pellucid; VP drainage with Delta (Medtronic) low pressure valve.



Case 4.

64 year old male patient facing important balance disorders, persistent vertigo, extremely difficultgait, urinary incontinence (and incipient stercoral), onset of Alzheimer disease (after neurological and psychiatricexaminations). It was inserted a Delta low pressure valve. The symptoms improved partially after3-4 months; the gait has become easier, sphincter disturbances have improved and the cognitive impairmentstill exist, but more tolerable.

Case 5.

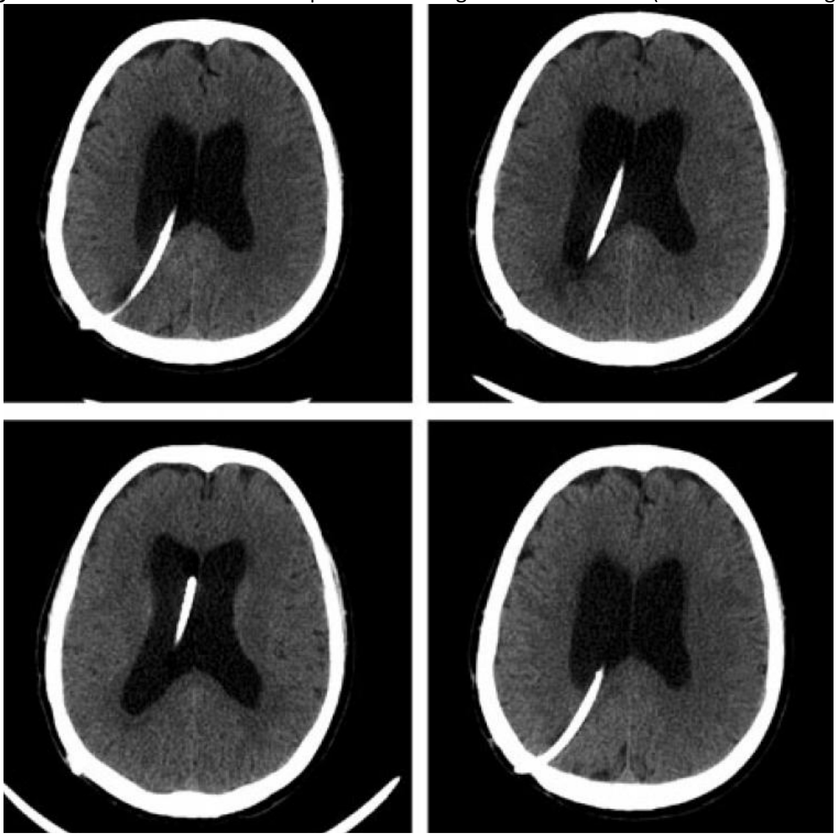
72 year old female patient hospitalized for walking difficulties, balance disorders, persistent vertigoand vomiting, dehydration. A central venous catheter was inserted into the right subclavian vein andVP drainage with a Delta low-pressure valve was inserted on the left side; intraoperative CSF pressure was15 mmHg.

Case 6.

66 year old male patient with memory disorders, impaired balance and gait disorders, cognitiveimpairment, urinary incontinence (occasionally), started over a year ago, with progressive evolution. Intraoperativemeasured ICP was 18 mmHg. Ventriculoperitoneal drainage with Holter low pressure valve wasinstalled. The

symptoms improved after one month with the complete remission of gait disorders, impairedbalance and sphincter disturbances.

Figure 5. The ventricular catheter placed inside right lateral ventricle (functional drainage).



CONCLUSIONS

The ventricular enlargement and the pressureon the frontal lobes are probably responsible for theoccurrence of cognitive disorders and dementia.

The pressure on the regulator centers of the sphincterfunctions located in the paracentral lobule maybe responsible for the urinary incontinence. Theventricular dilatation may compress the internalcapsule and secondary the pyramidal tract, responsiblefor the gait disorders, impaired balance and pyramidalsyndrome (4, 5, and 9).

To determine the surgical indication and to anticipatethe subsequent postoperative evolution, severalclinical and imaging criteria must be established:

• the obstructive causes for hydrocephalus must be excluded;it must be a communicating form;
• a periventricular hypodensity on CT scan or hyperintensityon T2 sequence (MRI) might be transependimar CSFresorption and might anticipate a favorable evolutionafter the shunt procedure;
• the rounding and symmetrical ballooning of the frontalhorns;
• dilated focal of convexity sulci may be revealed by imagingstudies and they are atypical CSF reservoirs whichsubsequently decrease after drainage and they should notbe confused with cortical atrophy; typical cortical atrophy(ex vacuo hydrocephalus) occurs frequently with Alzheimerdisease, and there is a limited response to ventriculoperitonealdrainage (11).

Figure 6. Preoperative MRI (T2 sequence) and postoperative CT scan; right ventricular catheter;prominent convexity sulci; incipient cortical atrophy

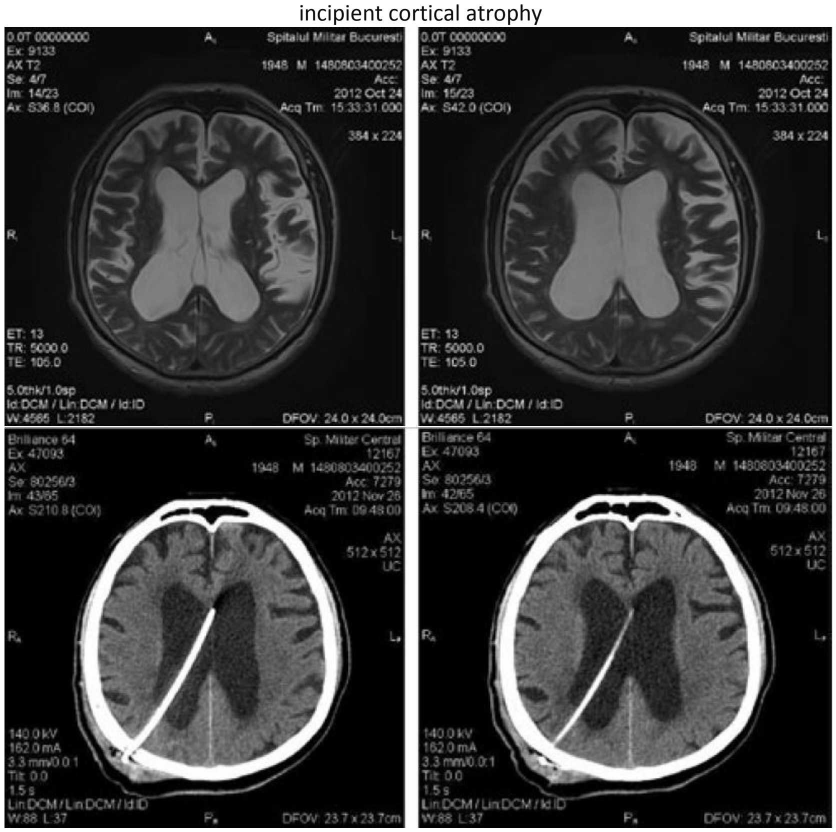


Figure 7. Preoperative and postoperative CT images; the catheter has been placed inside the left lateral ventricle because a central venous catheter was inserted into the right subclavian vein (functional drainage).

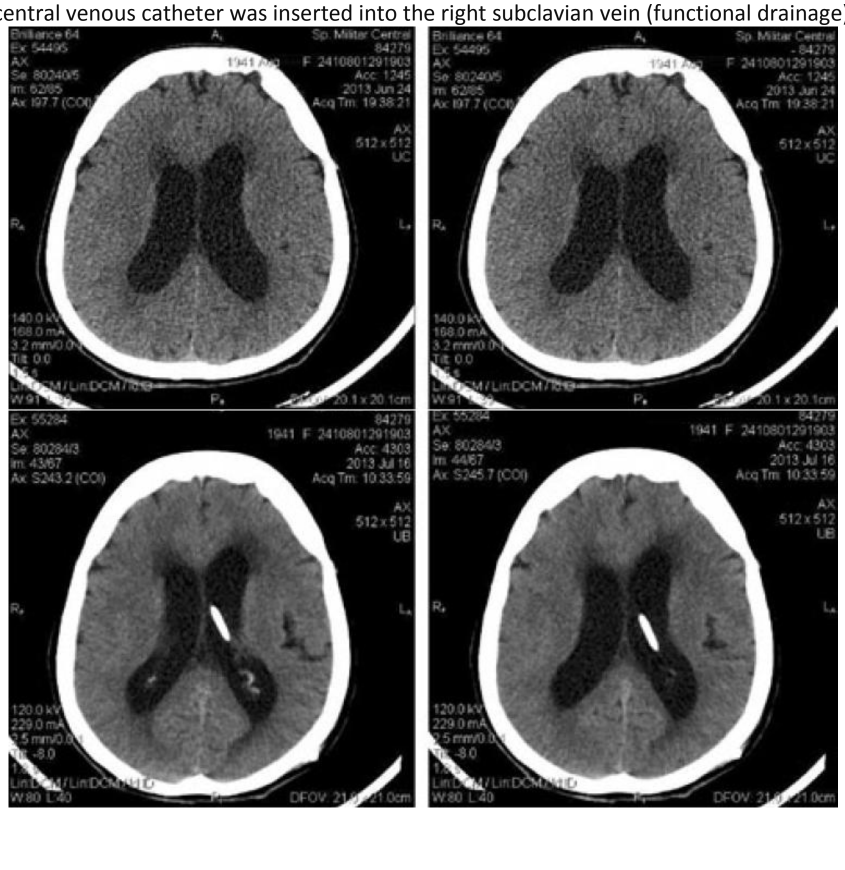
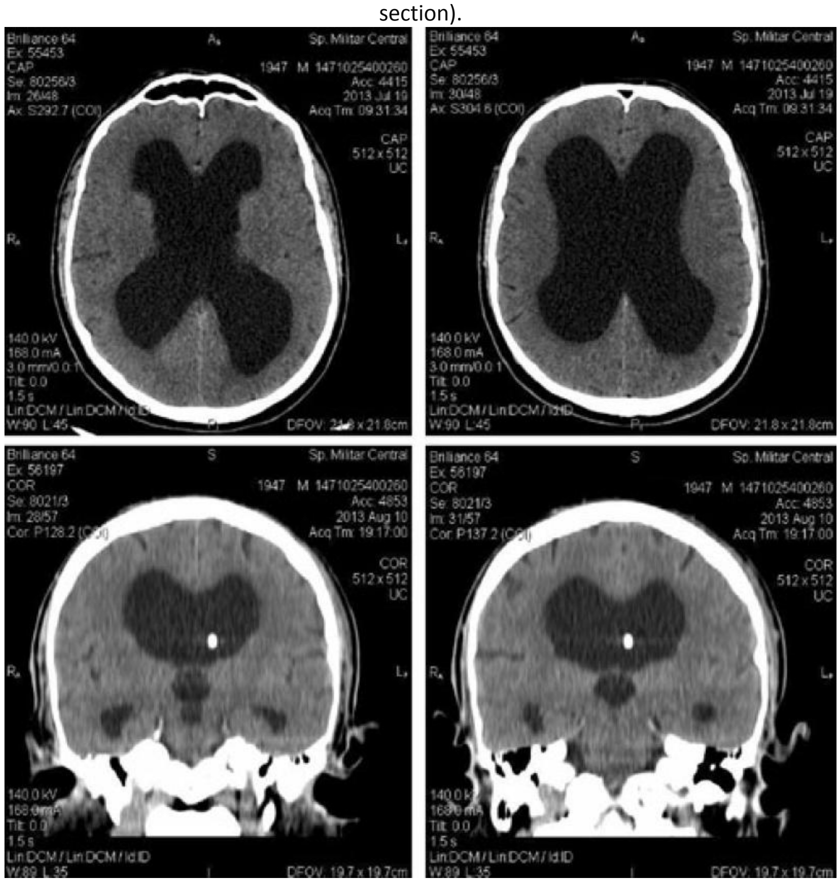


Figure 8. Important hydrocephalus with cortical sulcus persistent; catheter placed inside the left lateral ventricle (coronal section).



Maintaining good results depends on periodicmedical checks, immediate recognition and treatmentof complications (insufficient drainage shouldbe suspected first). There is no ideal method for thetreatment of hydrocephalus. None of the surgicaltechniques is perfect and no device gave total satisfaction.

The accelerated development of technology andthe practical experience in CSF intra- and extrathecaldrainage allow overcoming the current difficulties.

The world population undergoes a pronounced agingprocess, the number of people aged over 60, increasingto 400 million over the last 40 years. The ratio ofthe elderly population has been modified, representingover 50% in developed countries, with the Europeanzone being the aged (10).

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