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Cerebrospinal compensation in hydrocephalic children

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Abstract. One hundred and fifteen cases of hydrocephalus in children were analysed. Cerebrospinal compensatory reserve was assessed by a computerized, constant rate, lumbar infusion test. Head circumference and ventricular size were measured and a psychometric examination carried out. A classification of hydrocephalus based on resting cerebrospinal fluid pressure (CSFP) and resistance to cerebrospinal fluid outflow (RCSF) was introduced. Parameters of compensatory reserve were compared in atrophy (low CSFP, low RCSF), normal-pressure hydrocephalus (low CSFP, increased RCSF), non-communicating hydrocephalus (high CSFP, low RCSF) and acute hydrocephalus (high CSFP and increased RCSF). Significant differences were found between the factors describing compensatory ability in these groups. Sixty-two patients could be classified on the basis of resting CSFP and RCSF. Differentiation between the types of hydrocephalus was shown to be more accurate when all variables measured during the pressure-volume test were considered. The patterns of the time courses of CSFP during rate infusion tests in the different types of hydrocephalus are presented.

Key words: Hydrocephalus–Cerebrospinal fluid pressure–Cerebrospinal compensation–Infusion test

Introduction

It is still difficult to choose the optimal treatment for children with hydrocephalus [5, 8, 12, 14, 15, 18, 22]. Does knowledge of cerebrospinal fluid (CSF) dynamics help? Pressure-volume compensatory reserve is tested before shunting in many centres [3, 8, 11–13, 21]. Cerebrospinal fluid pressure (CSFP), resistance to CSF outflow (RCSF) and cerebrospinal compliance have all been measured for many years [2, 3, 11, 17–20], but debate continues as to their prognostic significance [2, 11, 18, 22]. We have used a computerized lumbar infusion test [7] to expedite the measurement of a selection of variables describing cerebrospinal fluid dynamics and have explored their usefulness in the characterization of compensatory reserve in 115 cases of children with ventricular dilatation. A characterization arrived at this way may give a rational basis for making a decision about shunting; this can be proved by statistical analysis of the results of shunting in different classification groups. With an average patient age of 3.5 years, conclusive follow-up will be possible in about 15 years time. This limits the usefulness of the classification introduced, but it can still be helpful in understanding the different mechanisms involving deficits in cerebrospinal compensation in paediatric hydrocephalus.

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Patients and methods

A group of 115 children with ventricular enlargement, confirmed by computed tomography (CT) and, in the majority, symptoms of so-called arrested hydrocephalus [10] were examined at the Neurosurgical Clinic, Children's Health Centre, Warsaw. This type of hydrocephalus is characterized by an arrest in the advance of pathological symptoms and is the most difficult type to assess when deciding on whether shunting is necessary. This decision is usually made on the basis of increased level of resistance to CSF reabsorption and increased resting CSF pressure. The role of the pressure-volume test, which provides information on cerebrospinal compensatory reserve, is important in this type of patient. No improvement after shunting can be expected if cerebrospinal compensation was not impaired before the shunt was put in place. Therefore, the test can help to avoid unnecessary shunting.

For the statistical analysis cerebrospinal compensatory reserve and a number of clinical variables were compared.

Cerebrospinal compensatory reserve

The procedure for the computerized lumbar infusion test has been described previously [21]. The lumbar CSF pressure (CSFP) signal was analysed by a computer system developed at the Warsaw University of Technology [7]. The following parameters were compared statistically: *average resting CSFP* ($CSFP_r$) during the 5-min period of recording before the infusion. The criteria for exact measurement

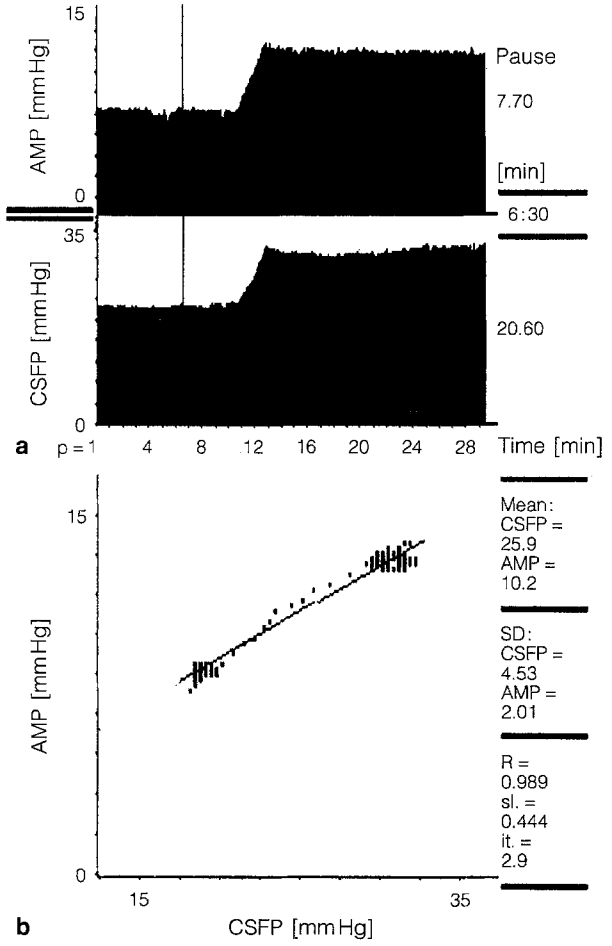

Fig. 1. **a** Time trends of amplitude (*AMP*) and mean CSF pressure (*CSFP*) recorded during lumbar infusion test (hydrocephalus in Dandy-Walker syndrome, classified into group III). Infusion rate started at 10 ml/min and was switched to 1 ml/min when cerebrospinal fluid pressure increased by 10 mmHg. *Abscissa:* time in minutes counted from the beginning of recording. Values of amplitude and pressure are calculated every 4 s. **b** Amplitude-pressure characteristic evaluated on the basic of the infusion test presented above. The linear regression line is drawn and its parameters are displayed on the graph

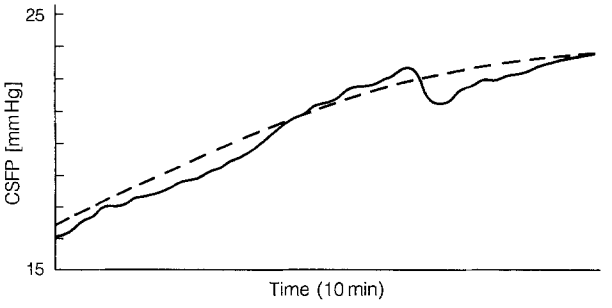

Fig. 2. Example of identification of the cerebrospinal compensatory model using minimization of the recorded increasing CSFP within the test and smooth theoretical curve. The roots mean square distance between the two curves is minimized with respect to unknown compensatory variables. This case (hydrocephalus following arachnoid cyst) was classified into group III. Note the characteristic notch of CSFP at the end of the test when the resistance of CSF outflow decreases due to widening of flow pathways on the brain convexity

of resting pressure are still unclear. The authors adopted their own definition of resting conditions as a period of stable CSFP (with no fluctuations slower than B waves). Additionally, the short-term correlation coefficient between amplitude and pressure was analysed. This describes how much spontaneous fluctuations in CSFP influence the changes in amplitude of CSF pulse pressure. In resting conditions when the net cerebrospinal volume is stable, this correlation is non-significant [6].

Pulse amplitude of CSFP in resting conditions (AMP_r) was calculated by the fast Fourier transform of CSFP evaluted every 4 s as the peak to peak of the first harmonic of the pulse wave [6, 7].

Gradient of the amplitude-pressure regression line (AMP/P) calculated on the basis of the trends of amplitude and pressure during the test (Fig. 1).

Resistance to CSF outflow (RCSF), pressure-volume index (PVI). Both values were calculated using the non-linear regression of recorded CSFP versus modelling curve [7]. The root mean square distance between these two curves (Fig. 2) is minimized during this procedure, with respect to unknown RCSF and PVI. The value of the minimal root mean square distance describes the total energy of pressure waves reflecting vasomotor activity, frequently increasing during the test as an evidence of the system's intolerance of the elevation in CSFP. Therefore, this parameter (distance) was analysed as a factor describing cerebrospinal vasogenic volume reserve.

CSR formation rate (I_{form}) estimated using the Davson's equation:

$$I_{form} = \frac{CSFP_r - p_{ss}}{RCSF}$$

p_{ss} is the pressure inside the sagittal sinuses. It is estimated as the point of intercept of the amplitude/pressure regression line and pressure axis.

Time constant of cerebrospinal space (τ), i.e. the product RCSF and the system's compliance (in resting conditions). The time constant describes the average time needed to evacuate fluid added from outside the system.

Cerebrospinal system compliance (C) is the inverse of the initial slope of increase in mean CSFP level just after the start of infusion.

Clinical variables

Age, head circumference (expressed as percentage of the deviation from the 50th percentile of norm), psychometric tests (Psyche-Catell 10 level scale: 6 denotes normal development) and ventricular dilatation ratio (calculated as arithmetical mean of bifrontal and bicaudal indices) from the CT scan.

These parameters were compared to each other using two-sample analysis in the Stratgraphics 5 system for IBM personal computers.

Results

The figures for the parameters describing the cerebrospinal compensatory reserve are presented in Table 1, which contains the mean values, medians and standard deviations for 115 cases. The patients were then classified to different groups. The criteria were low or high RCSF and the resting CSFP. Sixty-two of all 115 studied children were classified into four groups.

1. Group I: CSFP < 8 mm Hg, RCSF < 8 mm Hg/ml per min ($n=9$): Only enlarged ventricles suggest pathological changes in the brain. Low both RCSF and CSFP point out normal cerebrospinal compensation.

2. Group II: CSFP < 10 mm Hg, RCSF > 12 mm Hg/ml per min ($n=18$): Normal pressure hydrocephalus: low CSFP with the increased RCSF.

Table 1. Variables measured to assess cerebrospinal compensatory reserve in 115 children with dilated ventricles

	Mean	Median	SD
Resting CSFP (mmHg)	12.4	11.7	5.4
Resting pulse wave amplitude (mmHg)	3.56	2.9	2.56
Slope of AMP/P regression line (N.S.)	0.35	0.35	0.17
Resistance to CSF outflow (mmHg/ml per min)	10.1	9.5	5.13
Pressure-volume index (ml)	23	19	14
Estimated rate of CSF formation (ml/min)	1.20	0.728	1.2
Time constant (min)	19	10	26
Distance[a] (mmHg)	0.83	0.36	1.18
Compliance (ml/mmHg)	1.17	1.09	0.72

[a] Root mean square distance between real and predicted course of CSFP (see Fig. 2)

Table 2. Distribution of etiological factors within the four groups

	Group I (normal; %)	Group II (NPH; %)	Group III (obstructive; %)	Group IV (acute; %)
Congenital malformation	33	32	45	24
Birth trauma	66	54	55	0
CNS infection	11	21	17	81
Head injury	0	21	0	0

3. Group III: CSFP > 14 mm Hg, RCSF < 10 mm Hg/ml per min ($n = 28$): Normal RCSF with an increased CSFP in the lumbar subarachnoid space. Almost all the confirmed cases of obstructive hydrocephalus were seen in this group (as hydrocephalus following obstruction of foramina of Lushka and Magendie in Dandy-Walker syndrome (7 cases) [9], arachnoid cyst closing one or both foramina of Monroe (11 cases), etc.). These can be called "non-communicating" or "obstructive" types of hydrocephalus. A high CSFP in the lumbar subrachnoid space can be explained by the high-pressure gradient across the brain mantle, which causes a narrowing of the CSF flow routes on the brain convexity, thus producing an additional non-linear resistance to CSF outflow and increasing the CSFP in the subarachnoid space. During lumbar infusion there is a rise in CSFP in the lumbar channel and a decrease the transmantle pressure gradient. This results in a rapid decrease of resistance to CSF outflow – hence the normal RCSF measured in this group. Commonly, the characteristic notch on the mean-pressure curve (see Fig. 2) can be observed at the end of the test.

4. Group IV: CSFP > 14 mm Hg, RCSF > 14 mm Hg/ml per min ($n = 7$): These cases can be named "acute" because of the increased CSFP and RCSF that indicate the uncompensated stage of malresorptive hydrocephalus (i.e. causing intracranial hypertension). These are historical data from the years 1983–1984. We would now not recommend the test when CSFP is raised.

There were another 53 cases whose $CSFP_r$ and RCSF points lay within the border zones between groups (Fig. 3). The other variables relating to cerebrospinal dynamics were examined to determine if they could help in

Table 3. Results of pressure-volume study and clinical and CT findings in four groups (mean/SD)

	Group I (normal)	Group II (normal-pressure hydrocephalus)	Group III (obstructive)	Group IV (acute)
Resting $CSFP_r$ (mmHg)	5.7/1.6	7.0/1.9	18.1/4.1	15.3/2.5
Resistance to CSF outflow (mmHg/ml per min)	5.1/1.1	16.7/3.6	6.4/2.3	15.5/1.7
Resting pulse wave amplitude (mmHg)	3.8/3.6	2.2/1.1	5.3/3.3	3.5/1.5
Slope of AMP/P regression line (N.S.)	0.43/0.29	0.32/0.15	0.36/0.17	0.4/0.18
Pressure-volume index (ml)	19.4/13	20.3/16	41.3/41	16.2/6
Estimated rate of CSF formation (ml/min)	1.07/0.64	0.37/0.18	2.41/1.5	0.54/0.4
Time constant (min)	18.6/21	38/34	6.53/4	24/9
Distance[a] (mmHg)	0.438/0.23	0.89/0.38	0.97/0.52	1.69/0.7
Compliance (ml/mmHg)	1.82/0.8	1.47/0.9	1.27/0.9	0.97/0.2
Age (months)	42/41	24/17	47/51	51/63
Head circumference (% deviation from 50th percentile of norm)	7/7.6	8.2/6	7.4/6.5	7.9/6.8
Ventricular dilatation ratio	0.58/0.18	0.52/0.1	0.47/0.17	0.60/0.13
Psychomotor development (Psyche-Catell scale)	5.1/0.9	4.2/1.1	4.9/1.3	5/1

[a] Root mean square distance between real and predicted course of CSFP

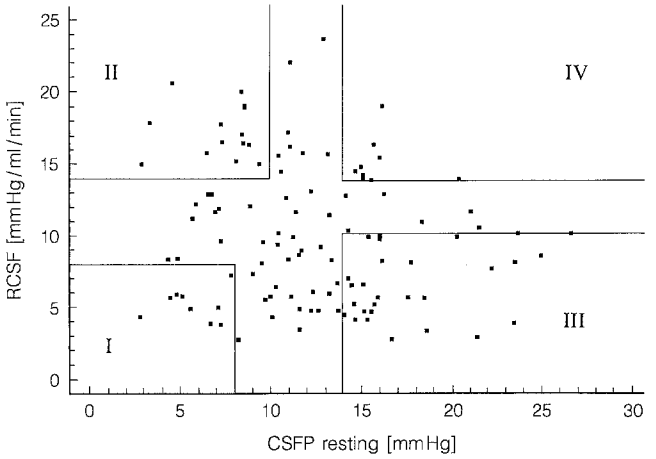

Fig. 3. One hundred and fifteen cases of hydrocephalus in children, expressed as points on resistance to CSF outflow (*RCSF*) vs resting CSFP. The four groups: I, normal; II, normal-pressure hydrocephalus; III, obstructive hydrocephalus; IV, acute or malresorptive hydrocephalus

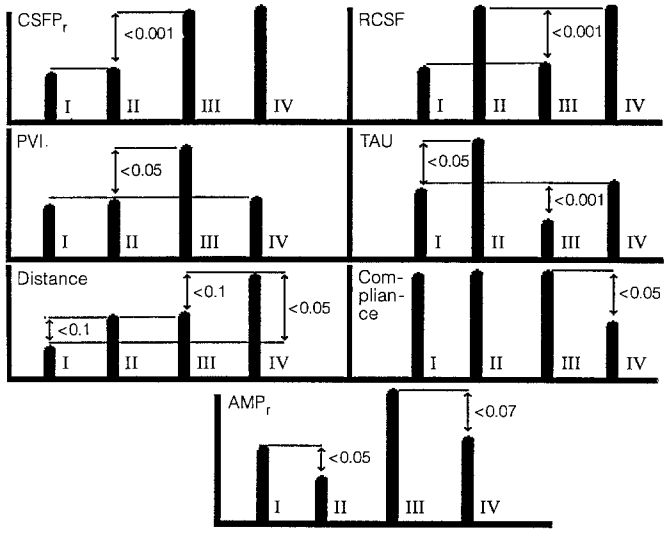

Fig. 4. Statistical differences between chosen compensatory variables in the four groups. *Black bars* represent the means in each groups; statistical significance of differences between means are indicated (two-sample test)

classifying these "uncertain" patients. This is important because patients in groups II, III and IV should in general be considered to have hydrocephalus that can be treated by shunting, while there is little prospect for improvement after surgery for patients in group I.

The distribution of etiological factors within groups discussed is shown in Table 2. The variables measuring cerebrospinal compensatory reserve (mean values and standard deviation) clinical variables and psychometric assessment calculated separately for each group are given in Table 3. A two-sample analysis of statistical significance was carried out. The results showing the variables that differ statistically in pairs are presented in Fig. 4.

A correlation between CSFP and RCSF outflow was found only in group IV (acute hydrocephalus): $r=0.81$; $\mathrm{RCSF}=0.53\cdot\mathrm{CSFR}+8.51$; $r<0.026$. In the remaining three groups, a significant correlation was found between the resistance and end-equilibrium CSFP reached during the infusion test.

There was no correlation between RCSF and PVI in any of the groups, which is different than the findings reported in adults [19].

Discussion

In group I no noticeable deficit in the compensatory reserve was found. The only pathological symptom was enlargement of the ventricles and accelerated head growth. The psychometric results remained normal. The lowest root mean square distance between the real and theoretical trends of CSFP was the only compensatory variable with a significant difference ($P<0.05$) from other three groups. This reflects that the rise of CSFP recorded during the infusion test in patients with normal cerebrospinal volume compensation is almost perfectly smooth without any of the vasomotor responses seen in other uncompensated forms of hydrocephalus [6, 16]. This group has the highest cerebrospinal compliance (although this is not statistically significant, it does not differ from that in the normal-pressure hydrocephalus group at the $P<0.1$ level).

Group II (normal-pressure hydrocephalus) had: the lowest resting pulse-wave amplitude; the longest cerebrospinal system time constant; the lowest (and nearest to the physiological norm) estimated rate of CSF formation (but not significantly different from that in the acute hydrocephalus group).

Group III (obstructive hydrocephalus) had: the highest pulse-wave amplitude; the shortest time constant; the highest PVI; the highest estimated rate of CSF formation.

Group IV (acute or malresorptive hydrocephalus) exhibited the highest root mean square distance between the real CSFP and theoretical trends. It also had the lowest cerebrospinal compliance. There were significant differences between this and group II in the lower time constant and higher pulse wave amplitude. To differentiate between the "acute" group (group IV) and group III, the PVI and time constant must be considered: in group IV PVI is significantly lower and the time constant greater.

The ventricular dilatation ratio and psychomotor development did not differ significantly between groups (the largest ventricular dilatation ratio was in group IV). The greatest head circumference was found in group II, but this, however, was not statistically significant. Such a homogeneous clinical picture shows that in arrested hydrocephalus differentiation between patients exhibiting different states of cerebrospinal compensation cannot be done precisely without the pressure-volume test.

The above results show that in addition to CSFP and RCSF the remaining cerebrospinal compensatory variables can explain differences between the types of hydrophalus. Normal-pressure hydrocephalus can be considered as the compensated stage of acute hydrocephalus [22]. It differs from hydrocephalus in which intracranial

hypertension develops (groups III and IV) only in variables reflecting a larger buffering capacity (i.e. lower CSFP, lower pulse wave amplitude, and longer cerebrospinal time constant).

Acute hydrocephalus is undoubtedly the stage where the raised RCSF is responsible for the increase in resting CSFP. That this group had the highest correlation between RCSF and CSFP therefore seems to be obvious. This group also shows the highest vasomotor activity during the infusion test, demonstrating low vascular compensatory reserve (i.e. the ability to compensate the changes in CSF volume by reciprocal shifts in cerebral blood volume) [16]. Group III, which contains several types of obstructive hydrocephalus, is characterized by sufficient dynamic compensatory ability manifested by the highest PVI. However, the highest pulse wave amplitude and the shortest time constant may suggest a contradictory conclusion. These support the hypothesis of dynamic compensation by the shifts of the brain mantle, due to the changes in the ventriculo-subarachnoid pressure gradient. Because this "dynamic" compensatory volume reserve is very limited, the cerebrospinal time constant is shortest. During the gradual increase in CSFP in the lumbar subarachnoid space, narrowed pathways of CSF circulation on brain convexity are widened and CSFP is then stabilized by CSF reabsorption. Davson's equation describing the static compensation is strongly non-linear in this group, which can be detected using advanced forms of system identification in the computerized infusion test program [7].

Differentiation between the three groups of "active" hydrocephalus and the atrophic group (group I) is not difficult if we take into account all compensatory variables. The most important problem, that of distinguishing between atrophy and normal-pressure hydrocephalus, can be solved by considering such variables as the root mean square of the distance between the real and predicted trends of CSFP recorded during the test, pulse wave amplitude or estimated CSF formation rate.

To validate the above characterization, a new classification was produced on the basis of CSFR, RCSF and remaining compensatory parameters. The border limits for CSFP and RCSF were changed to cause the different groups to overlap each other on the CSFP/RCSF plane.

Class I: Normal: CSFP <12, RCSF <12, distance $<$ median

Class II: Normal-pressure hydrocephalus: CSFR <12, RCSF >10, $\tau >$ median, $AMP_r <$ median

Class III: Non-communicating hydrocephalus: CSFP >10, RCSF <12, $\tau <$ median, $AMP_r >$ median

Class IV: Acute hydrocephalus: CSFP >10, RCSF >10, distance $>$ median, PVI $<$ median

where "median" denotes the median values for distribution of each parameter in 115 cases (these values are given in Table 1).

These groups contain more "uncertain" cases previously lying in the border zones–see Fig. 2 (I, 24; II, 28; III, 29; IV, 15). It should be noted that the differences in mean values of CSFP and RCSF between these classes were not affected and were statistically significant, as in the first classification.

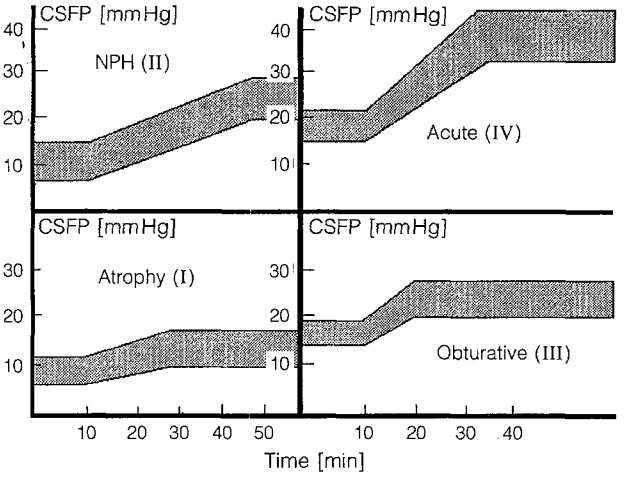

Fig. 5. Differences between courses of increasing CSFP during infusion tests in the discussed four groups. *Shaded area* shows upper and lower quartile limits of CSFP for each group

Finally, the differences between end-equilibrium levels of CSFP reached during constant-rate (1 ml/min) lumbar infusion were also analysed. This enabled the patterns of time coursed of mean CSFP during the constant rate infusion test to be introduced in each group separately.

In both the atrophic and the normal pressure hydrocephalus groups CSFP started from the same level (8 ± 2.7 mm Hg). The end equilibrium level in atrophy was 16 ± 5 mm Hg and in normal-pressure hydrocephalus 27 ± 8 mmHg. In group III (obstructive or hydrocephalus) CSFP started from 16 mm Hg $\pm$ mm Hg and reached 27 ± 8 mm Hg (the same as in group III). In the acute group it started from the same level as in group III, but reached the level 41 ± 8 mm Hg (see Fig. 5). Different gradients of increase in mean CSFP reflect the differences discussed between cerebrospinal compliances and time constants in the various types of hydrocephalus.

In conclusion, the classification of hydrocephalus in children on the basis of cerebrospinal compensatory variables explains the differences in intracranial biomechanical properties in normal pressure, obstructive, malresorptive hydrocephalus and brain atrophy. Resistance to CSF outflow and mean resting CSFP alone do not permit precise classification of all cases. Variables such as pressure-volume index, cerebrospinal compliance, time constant, resting amplitude of pulse wave make the differentiation more reliable. This classification is clinically useful in helping to avoid unnecessary shunting: cases classified into the atrophic group have little chance of benefiting from shunt insertion.

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