Classification of Ventriculomegaly in Adults: A Cluster Analysis

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ABSTRACT

Object

Chronic ventriculomegaly in the absence of raised intracranial pressure (ICP) is a known entity in adult hydrocephalus practice. The natural history and indication for treatment is however, poorly defined. A highly heterogeneous group, some adults with ventriculomegaly are asymptomatic, whilst others have life-threatening deteriorations. We hypothesise that the various presentations can be sub-typed and represent different stages of decompensation. We perform a cluster analysis on a cohort of patients with chronic ventriculomegaly with the aim to elucidate typical clinical characteristics and outcomes in chronic ventriculomegaly in adults.

Method

Data was collected from 79 patients referred to a single centre with chronic ventriculomegaly, including demographics, presenting symptoms, 24-hour intracranial pressure monitoring (24-hour ICPM). We perform a statistical cluster analysis to determine the presence of sub-groups.

Results

Four main subgroups and one highly dissimilar group were identified. Patients with ventriculomegaly commonly have a perinatal event followed by four main presentations: (1) incidental ventriculomegaly with or without headache, (2) highly symptomatic presentation (including reduced consciousness) and raised ICP (3) early-presenting with symptoms of headache and nausea (with abnormal pulsatility) and (4) late-presenting with features common to NPH. Each symptomatic group has characteristic radiological features, ICPM and responses to treatment.

Conclusion

Cluster analysis has identified sub-groups of adult patients with ventriculomegaly. Such groups may represent various degrees of decompensation. Surgical interventions may not be equally effective across the sub-groups, presenting an avenue for further research. The identified sub-types provide further insight into the natural history of this lesser studied form of hydrocephalus.

Key words: Long-standing overt ventriculomegaly (LOVA), ventriculomegaly, classifications, cluster analysis, hydrocephalus, endoscopic third ventriculostomy (ETV), Ventriculoperitoneal shunt (VP shunt)**.**

INTRODUCTION

Chronic ventriculomegaly in the absence of raised intracranial pressure (ICP) is a known entity in adult hydrocephalus practice (figure 1). The improved availability of brain imaging has meant that more patients are discovered to have large ventricles. Likely to evolve in infancy, it tends to present in adulthood with a range symptoms and poorly defined natural history[15].

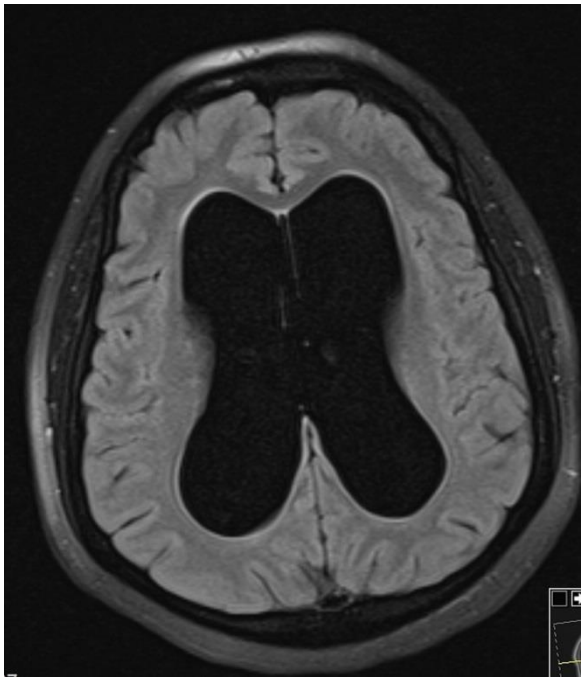

Figure 1. MR showing typical appearance of ventriculomegaly in LOVA

There are various terminologies for ventriculomegaly in adults, including long standing overt ventriculomegaly in adults (LOVA), aqueduct stenosis with hydrocephalus, adults with previously untreated congenital hydrocephalus (APUCH), syndrome of hydrocephalus in young and middle-aged adults (SHYMA), chronic congenital hydrocephalus, arrested hydrocephalus, or compensated hydrocephalus[2,7,15].

This group is a highly heterogeneous making the management of this condition a challenge. Patients may be present at any age, can be asymptomatic or present in extremis in a vegetative state[8,12,15]. Radiological features are also variable, with some patients having aqueduct stenosis and others not[9]. Due absence of any randomised controlled trials; it is unclear what the optimum management for LOVA is, with options including endoscopic third ventriculostomy (ETV), ventriculoperitoneal (VP) shunt, and even dural venous sinus stenting[8-10,12,15,17].

We hypothesise that the various symptomatic presentations represent different stages of decompensation and that identification of the various stages will aid management strategies. We perform a cluster analysis on a cohort of patients with chronic ventriculomegaly with the aim to elucidate typical clinical characteristics and outcomes in chronic ventriculomegaly in adults.

METHODS

Study design

We perform a retrospective cohort study and cluster analysis. Radiological and clinical data was recorded prospectively and analysed retrospectively.

Inclusion criteria

Potentially eligible patients were consecutively identified from the single centre's records and had been referred to this single centre between December 2012 to May 2016. Referrals including both incidental and symptomatic ventriculomegaly. Inclusion criteria included adults (patients ages 18 years or more), who underwent parenchymal intracranial pressure (ICP) monitoring with confirmed radiological evidence of ventriculomegaly.

Exclusion criteria

Patients with acute hydrocephalus (evident by periventricular oedema and return to normal ventricular dimensions post CSF diversion), normal pressure hydrocephalus (defined by the international guidelines for probable INPH) or those with radiological evidence of NPH (''DESH'' Sign) were excluded[7,17]. Patients with ventriculomegaly in the absence of symptoms did not under the invasive test of ICP monitoring.

Clinical evaluation

Basic demographic data on gender and age was recorded at the point of referral. Glasgow Coma Score (GCS), and the categorical presence of symptoms (cognitive deficit, gait, incontinence, headache, dysphagia, fatigue, visual deterioration, nausea, balance, and epileptic seizures) were confirmed at a minimum of two clinic appointments with a neurosurgical or neurological consultant. Fundoscopy (and if possible retinal photography) was performed to examine for papilloedema. Clinical records were reviewed for the presence of obstructive sleep apnoea (OSA) and perinatal injury.

Radiological investigations

Consistent presence of ventriculomegaly, defined by enlarged lateral ventricles (Evans' index greater than 0.3) measured at the point of foramen of Monroe and without features of a) acute raised ICP (i.e. should not have evidence of periventricular oedema) and without b) generalised cortical atrophy on MR images or CT (exemplified by figure 1)[13]. A consultant

neuroradiologist and two clinicians determined ventriculomegaly independently. In one instance did the reports not concur. In this instance the majority opinion prevailed. This was repeated to confirm the presence of tri-ventriculomegaly or Pan-ventriculomegaly, The mean Evan's index (EI), calculated by two independent clinicians, is reported.

24-hour ICPM

24-hour ICPM using intraparenchymal intracranial monitoring devices (Spiegelberg GmbH & Co. KG, Hamburg) collected minute-by-minute data determined median ICP (during day and night) and pulsatility. Median ICP (during day and night) was recorded. Median pulsatility (difference between systolic and diastolic ICP values) greater than 4mmHg was considered raised[1]. ICP data was processed using Excel (Microsoft ©, Reading). All ICPM data was analysed prospectively. ICP is presented as mmHg (1mmHg = 1.36cmH_2O).

Intervention and outcome

Clinical records were reviewed for follow-up (clinic), intervention and grouped into 'no intervention', medical management, surgical intervention (with endoscopic third ventriculostomy or ventricular shunting). A procedure was deemed successful if, at the time of follow-up the patient had a clinical improvement documented (subjectively reported resolution in any presenting symptom) and stable radiological features. Any requirement for further surgical intervention was recorded. Differences in intervention and outcome between the observed groups were studied post cluster-analysis.

Statistical analysis

Cluster analysis was performed using the above demographic (gender and age), radiological (tri-ventriculomegaly vs. pan-ventriculomegaly and Evan's index[13]) and clinical data (GCS, symptoms, presence of OSA and perinatal injury). These variables underwent hierarchical agglomerative clustering with complete linkage. Gower's dissimilarity index (for continuous and categorical data) was used to measure the similarity and dissimilarity between the patients[5]. The results are visualised using dendrograms. Post-hoc comparisons to determine significance of clusters included analysis of variance (ANOVA) (for continuous variables) and chi-square tests (for the categorical variables). Significant level was set at alpha = 0.05 and tests were two tailed. Statistical analyses were done with programme R© Version 3.2.4 (R Foundation, Vienna).

RESULTS

Demographics and patient characteristics

A total of 508 patients with ventriculomegaly were identified, of which 270 were excluded as having acute hydrocephalus, and 114 excluded with a diagnosis of NPH and 45 patients had radiological features of ventriculomegaly (n=38), but had no clinical indication ICP monitoring,

or were to unwell and required immediate CSF diversion without ICP monitoring (n=6). A total of 79 with chronic ventriculomegaly patients (43 F: 36 M) with a follow up 69.7 ± 42.4 months (mean ± SD) were identified. A summary of patient demographics and characteristics is shown in Table 1.

Table 1. Demographic, radiological and clinical characteristics (Mean±SD or n%)	
36: 43	Sex (Female: Male)
41.9±16.9	Age (years) at referral
69.7±42.4	Follow up (months)
Clinical features	**n (%)**
24 (30%)	Cognitive deficit
31 (39%)	Gait
16 (20%)	Incontinence
60 (76%)	Headaches
2 (3%)	Dysphagia
13 (16%)	Fatigue
21 (27%)	Vision
12 (15%)	Nausea
13 (16%)	Balance
17 (22%)	Seizures
3 (4%)	Obstructive sleep apnoea
15 (19%)	History of pre-natal injury
Radiological and ICPM	**Mean±SD or n (%)**
68 (86%)	Triventriculomegaly
4 (5%)	Lateral horn enlargement
7 (9%)	Panventriculomegaly
62 (78%)	Sellar enlargement

0.50±0.11	Evan's Index
5.34±4.41	24-hour ICPM (mmHg)

The most common symptoms included headaches (76% of cases), gait disturbances (39%) cognitive deficits (30%, including mild learning difficulties, reduced IQ or memory impairment) and visual disturbances (27%, including retinopathy, refractory errors and strabismus and only 1 case of papilloedema which was unilateral).

Sixty-seven (86.0%) had a triventricular hydrocephalus, four (5.1%) had a degree of fourth ventricular enlargement and seven (8.9%) had lateral horn enlargement only. Sixty-two (78%) patients had evidence of sellar enlargement on and the mean Evan's Index was 0.50 ± 0.11.

Cluster analysis

Five patient clusters with common features were identified, with the fifth being highly dissimilar ($p<0.005$) (figure 2).

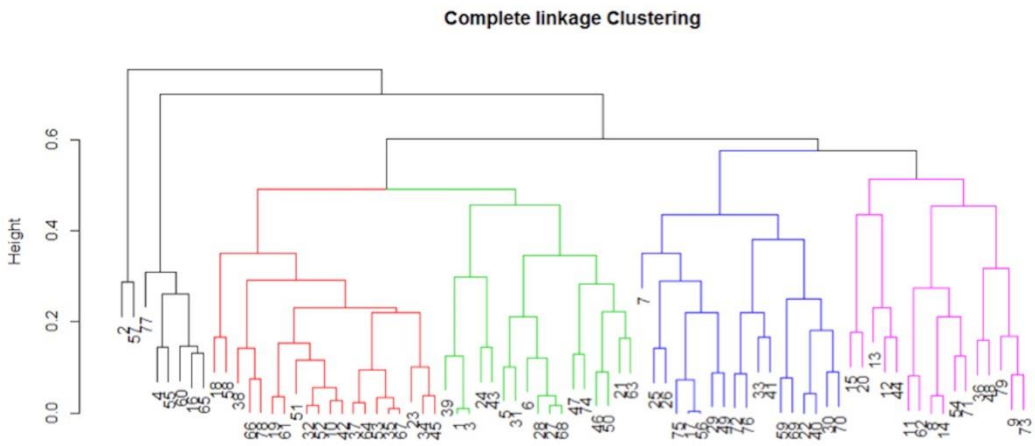

Figure 2. Hierarchical clustering showing 4 major clusters and a 5th highly dissimilar cluster

The key features that differentiated the sub-groups were gender, age, EI, 24-hour ICPM, and the presence of gait abnormalities, incontinence, headaches, fatigue, visual symptoms and nausea. Table 2 presents the major clusters and the variables within them.

Table 2. Summary statistics of variables for the main clusters observed						
p-value	Cluster 5 (n=8)	Cluster 4 (n=16)	Cluster 3 (n=20)	Cluster 2 (n=18)	Cluster 1 (n=17)	Variables

0.0001	2	15	12	3	8	Female
0.0001	40.0±14.7	59.1±17.4	28.9±10.7	48.3±15.6	39.8±11.3	Age
0.003	0.44±0.12	0.55±0.12	0.55±0.07	0.47±0.11	0.46±0.08	Evan's Index (EI)
0.010	4.2±0.80	1.71±0.37	4.0±0.96	14.0±2.45	3.10±0.21	24-hour ICPM (mmHg)
0.070	4	8	2	5	5	Cognition
0.0001	4	8	0	16	3	Gait
0.001	1	5	1	9	0	Incontinence
0.019	2	10	20	13	15	Headache
0.195	2	0	0	0	0	Dysphagia
0.005	2	7	1	1	2	Fatigue
0.0001	0	2	0	16	3	Vision
0.018	2	0	6	1	3	Nausea
0.141	3	5	1	2	2	Balance
0.162	7	2	5	0	3	Epilepsy
0.394	2	0	0	1	0	OSA
0.406	4	2	5	1	3	Preterm post-natal Injury

Categorical variables are presented as counts; continuous variables are presented as mean±SD. Bold font represents significant results.

The key features of each of the 5 clusters are summarised below:

Cluster 1 (n=17): This group had the fewest symptoms and normal ICP data, patients with ventriculomegaly. Patients in this sub-group were a mean age of 39.8 yrs. The predominant reason for patients in cluster 1 to undergo imaging was to investigate severe headaches refractory to medical management. This cluster represents those with incidental ventriculomegaly.

Cluster 2 (n=18): Patients in cluster 2 had raised ICP and were highly symptomatic, often presenting acutely. There were low rates of revision for both treatment modalities ETV (n=6) or VP shunt (n=12), with one patient requiring revision of their ETV to a shunt (due to persistent symptoms).

Cluster 3 (n=20): Indentified patients who on average were younger and had the largest ventricles. A notable finding was that 100% of patients in this group suffered with headaches. This group had normal pressure on ICP monitoring but impaired pulsatility. Two out of the six ETV's required revision and one of the 13 primary shunt insertions required revision.

Cluster 4 (n=16): Patients in this group presented at an older age and symptomatically mimicked some of the symptoms seen in normal pressure hydrocephalus. A severe headache (8/10 on visual analogue scale) was the predominant feature in ten of the 16 patients. Only three patients exhibited the full triad of NPH-symptoms and no patient had the typical 'magnetic gait' seen in NPH.

This group of patients improved after CSF diversion, but were prone to overdrainage (n=4). Two patients required shunt ligation followed by subsequent un-ligation adjustment of the valve to a high setting of 20mmHg. Two were conservatively managed by increasing the shunt valve setting from 5mmHg to 20mmHg. Following resolution of the subdural, all four patients subsequently underwent gradually decreasing of the valve setting from 20mmHg to 5mmHg (n=2), 7mmHg (n=1) and 12mmHg (n=1) until symptom resolution.

Cluster 5 (n=8): This was a highly dissimilar group to the other clusters and represented patients with panventriculomegaly. This group had a range of symptoms and were successfully managed with a VP shunt.

Intervention and outcome

Table 3 summarises the intervention and outcomes in each group. Cerebrospinal fluid (CSF) diversion (via ETV or VP shunt) resulted in symptom resolution in 75% of patients. Over-drainage complications (subdural haematoma's) occurred in four patients in cluster 4. Three patients in cluster 1 had their headaches effectively managed with acetazolamide (250mg twice daily).

Table 3. Definitive Interventions and outcomes presented per cluster at the time of follow-up

Total (n=79)	Cluster 5 (n=8)	Cluster 4 (n=16)	Cluster 3 (n=20)	Cluster 2 (n=18)	Cluster 1 (n=17)	Variables

26 (33%)	4	7	1	0	14	No intervention
3 (4%)	0	0	0	0	3	Acetazolamide
12 (15%)	0	0	6	6	0	ETV
3/12 (25%)	0	0	2**	1*	0	ETV revision
38 (48%)	4	9	13	12	0	Shunt
4/38 (11%)	1	2***	1	0	0	Shunt revision
4 (5%)	0	5	0	0	0	Subdural
59 (75%)	4	8	15	18	14	Symptom resolution (and stable at follow-up)

ETV = endoscopic third ventriculostomy
*Revised to shunt
** One required shunt surgery and one repeat endoscopy
*** Both due to subdural haematomas post operatively

Complications

There were no complications from performing 24-hour ICPM in this group of patients over this time period.

DISCUSSION

Overall the patients' symptoms and radiological features are classically associated with a diagnosis of LOVA[15]. Shizuo Oi et al provide a diagnostic criteria for LOVA[15]. This criteria includes three main features; 1) overt ventriculomegaly involving the lateral & criteria third ventricles with obliterated cortical sulci on CT/MR imaging (as shown in figure 1, 2) clinical symptoms include macrocephaly with or without subnormal IQ, headaches, dementia, gait disturbance, urinary incontinence, vegetative state, akinetic mutism, apathetic consciousness, and parkinsonism 3) neuroimages with expanded or destroyed sella turcica[15].

Despite this definition, there appears to be a wide variation in presentation, age and clinical management of LOVA. We find that patients with ventriculomegaly decompensate (or present) with characteristic symptoms at various ages. The different symptomatic subgroups

may represent the various degrees of decompensation in chronic hydrocephalus, possibly owing to brain atrophy and changes in compliance over the decades. The distinct phenotypes (or clusters) of chronic ventriculomegaly in adults have distinguishing demographics, symptoms, radiology, ICPM and outcomes.

Cluster 1: Incidental ventriculomegaly (normal ICP)

This group represented 21.5% (n=17) of the patients with ventriculomegaly. Patients in this sub-group were a mean age of 39.8 yrs, with an equal representation of female and male patients (8:9 F:M). Overall they have fewer symptoms than the other groups, but suffered from headaches requiring investigation with ICP monitoring (n=15) and mild cognitive deficits (including dyslexia and mildly reduced IQ observed in the 3 patients who had undergone formal testing).

The predominant reason for patients in cluster 1 to undergo imaging was to investigate headaches, or as part of their epilepsy work-up, or for an unrelated reason (e.g. in the emergency department). This group had a mean EI of 0.46 and normal mean 24-hour ICP. One interesting radiological features that may account for the lack of symptoms in cluster 1 is the presence of a spontaneous third ventriculostomy (STV), observed in 6 (who had adequate MR imaging to detect it) (figure 3 A: Sagittal sequence showing defect in the floor of the third ventricle, B: CSF flow sequences demonstrating flow across the floor of third ventricle). The management of these patients consisted of annual surveillance in clinic (either neurology or neurosurgical). Three had their headaches managed with acetazolamide (250mg twice daily), whilst the rest had standard analgesia.

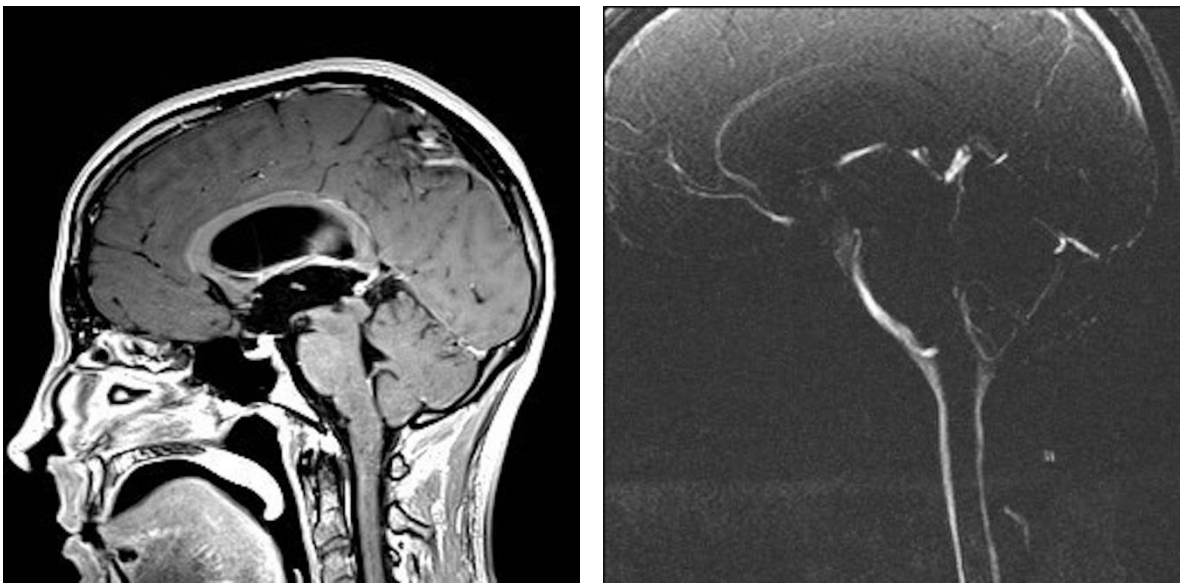

Figure 3. Spontaneous third ventriculostomy (STV) on MR imaging. A: Sagittal sequence (inverse) showing defect in the floor of the third ventricle, B: CSF flow sequences demonstrating flow across the floor of third ventricle in same patient.

Cluster 2: Decompensated ventriculomegaly (raised ICP)

Cluster 2 represented 23% (n=18) of the patients with ventriculomegaly and typically presented acutely as middle-aged adults. Interestingly this group had a disproportionate number of males (3:15 F:M ratio). The group was highly symptomatic with 89% having gait disturbances and visual disturbances, 72% suffering from headaches, 50% suffering from urinary incontinence and over a quarter suffered from a cognitive deficit. One of the 18 in cluster 2 had papilloedema However 'phenotypically cluster 2 type' patients had acute papilloedema bilaterally and acute GCS deterioration, but were excluded from this study as they needed immediate CSF diversion without ICP monitoring. These six patients probably represent the most extreme deterioration that can occur within this cluster.

Whilst their ventricles were large on MR (EI 0.47), following treatment their ventricles remained large and without periventricular interstitial oedema, suggesting this ventriculomegaly was chronic and had decompensated. Six patients in this group had aqueduct stenosis with no or very minimal flow on formal CSF flow studies. Whilst symptomatic this group had raised ICP's averaging 14.0mmHg.

The STV seen in some patients in cluster 1 may have protected against the eventual decompensation resulting in cluster 2 type LOVA. Increased thickness of the floor of the third ventricle may have prevented this protective STV from occurring. The decompensation seen in cluster 2 appears to take at least 3-4 decades to occur (assuming an initial insult in infancy), suggesting an ongoing maturation of the ventricular system, with reduced compliance and reduced ability to compensate over time.

Radiologically this group was very similar to cluster 1, highlighting the importance 24-hour ICPM to guide management. CSF diversion was performed in all 18 patients. The majority had a VP shunt (66%, n=12) and 6 (33%) had an ETV (the six with aqueduct stenosis). One ETV was subsequently revised to a shunt (due to persistent symptoms) 2 years later.

Cluster 3: Early presenting ventriculomegaly (normal ICP and impaired pulsatility)

The third cluster (n=20) notably presented in their second decade of life with headaches (100%) and nausea (30%) with an almost equal representation of female and male patients (12:8 F:M).

Whilst 24-hour ICPM was typically within a normal range (mean 4.0mmHg), abnormal pulsatility was common (mean 5.2mmHg).

Five patients within this group had a known periventricular haemorrhage associated with pre-term birth. A significant injury to the germinal matrix in the preterm post-natal period may explain why this group had particularly large ventricles (mean EI of 0.55). Whilst some of

these patients had aqueduct stenosis, they all had a degree of flow via the aqueduct on MR CSF flow studies.

The majority of patients in this group had a shunt (n=13). Six patients had ETVs. Two out of the six ETV's required revision, however; these had both been performed during adolescence and the patients now re-presented in their second decade of life. One of the shunts was revised due to proximal occlusion. Fifteen of the 20 had symptom resolution, but 5 patients had persistent symptoms including fatigue (n=1), cognitive deficit (n=1) and headaches (n=3).

The consistent finding of headache as presenting complaint was common. Such headaches in the presence of abnormal pulsatility may represent the beginning of a decompensation process. The presence of CSF flow may have protected this group against the severe decompensation seen in cluster 2.

Cluster 4: Late ventriculomegaly

The fourth cluster (n=16) presented in the fifth or sixth decade of life and symptoms mimic those seen in normal pressure hydrocephalus (this group could be referred to as 'NPH-like) including cognitive deficits (50%), gait disturbance (50%), balance disturbances (31%) and urinary incontinence (31%)[16]. The headache presenting in this group was the main complaint however.

The indications for ICP monitoring in this group were mixed. Ten of patients would ordinarily have undergone lumbar drainage for investigation of possible NPH (as per the international guidelines)[16]. However, due to contraindications (aqueduct stenosis, spinal canal stenosis, severe spinal deformity or previous arachoiditis in lumbar spine), ICP monitoring was undertaken to assist with diagnosis[4].

A subset of patients (n=6) in this group underwent ICP monitoring because they did not exhibit the typical triad of symptoms. None of these patients exhibited the characteristic magnetic gait of NPH.

This group was most commonly seen in women (ratio of 15:1 F:M). It is interesting to note that the late presenting group was predominately female, and the most highly symptomatic group (cluster 2) was predominately male. This may suggest that female gender has a protective effect preventing early severe decompensation.

The EI in cluster 4 was similar to cluster 3, being particularly large at 0.55. Similar to NPH, the 24-hour ICPM was normal (1.71mmHg). However, pulsatility in this group was predominately normal (mean 2.4mmHg), a feature that is typically abnormal in NPH[4]. The mean age 59.1 years, slightly younger than the mean age commonly seen in NPH[4].

One important distinguishing feature in this group is potential for of subdural haeamatoma's (SDH) to occur following insertion of a VP shunt. This is likely owing to the fact many patients had reduced compliance on their ICP monitoring (which may indicate that these patients did have raised ICP or abnormal pulsatility at some stage). One approach to preventing SDH in this group may include the use of a programmable valve and anti-siphon device, set at a high pressure (such as 18mmHg) initially, with a very slow reduction in the pressure over a period of months, titrated to symptom control.

Cluster 5: Panventriculomegaly

A fifth, smaller and more distant cluster was identified. Notably this sub-group had the presence of a panventriculomegaly. Symptomatically, the group had gait disturbance (50%) and cognitive deficits (50%) and a high incidence of epilepsy (88%) and pre-natal injury (50%), similar to patients in cluster 3 and in part 4. Some patients in this group also had dysphagia secondary to the enlarged fourth ventricle. All patients within this group were successfully managed with a VP shunt, with one patient requiring revision due to persistent symptoms.

Kageyama et al. reported cases of panventriculomegaly (with a wide foramen of Magendie and large cisterna magna) having similar symptoms to NPH (gait disturbances and cognitive dysfunction) and had hypothesised that such patients were a subtype of congenital hydrocephalus[11]. Due to the presence of panventriculomegaly, we consider this group a separate entity to the four groups above.

Management

There remain differences in opinion regarding the optimal management for LOVA, with overall favourable outcomes for ETV [9,10,12,15,17]. A recent paper by P.K. Eide concurs with our findings that LOVA is likely to have various subgroups with differing response to management. Eide found that those with normal ICP but abnormal pulsatility (Cluster 3 in our analysis) respond well clinically to CSF diversion[3].

Knowledge of CSF absorption and production may help select which patients would respond to ETV and to shunting[4]. Cluster 1, the group with incidental ventriculomegaly, is the only group that was not managed surgically. This may be due to either the presence of a spontaneous third ventriculostomy (described above) and may also be due to normal CSF absorption[4]. We advocate monitoring this group, as such patients may progress to becoming symptomatic.

Cluster 2 responded equally well to ETV and shunting and therefore may have an intraventricular CSF absorption failure. However, given the low numbers, it is not possible to

determine which was the superior treatment modality. As mentioned above, patients with LOVA are susceptible to over-drainage, and therefore should have shunts inserted with adjustable valves on a high-pressure setting that can be gradually titrated down.

Ventriculomegaly covers an age spectrum and presentation stage may dependent on the speed of development in CSF disturbances, or varying degree's of compenation. Long-term ventriculomegaly may induce future neurological changes various types or 'stages' of LOVA may evolve to another type (e.g. Cluster 1 may present as Cluster 4, later in life).

We present a flowchart (figure 4) to summarise the key clinical features, radiological and ICP findings of the four main subtypes identified in this study, with suggested management options. We emphasize the potential for various types to progress to other types.

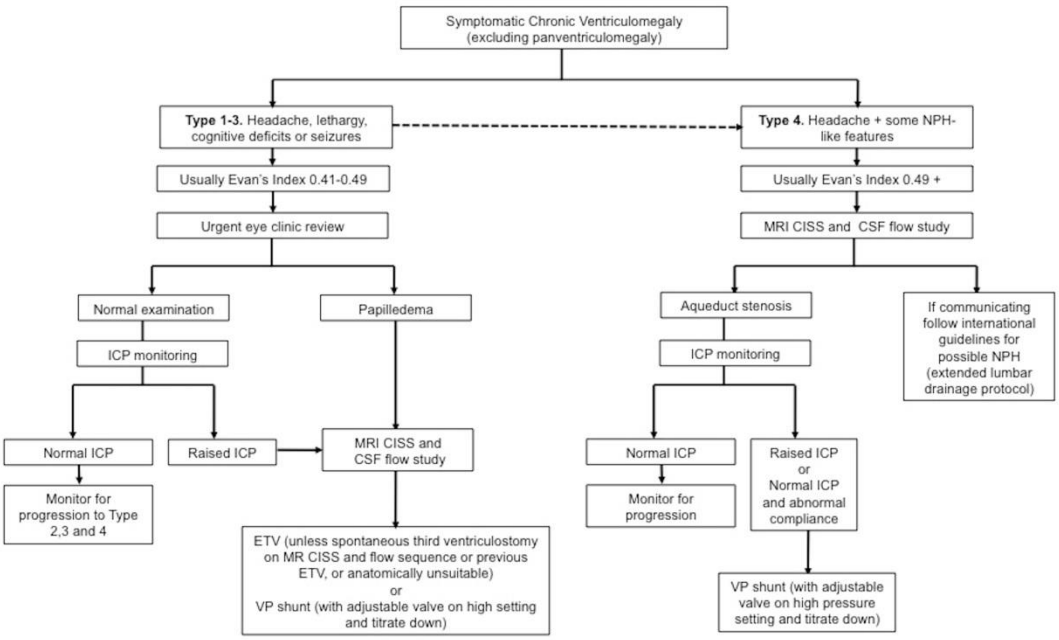

Figure 4. Flowchart summary of the four main subtypes clinical features and suggested management options (excluding panventriculomegaly). We emphasize the potential for various types to progress to other types (progression represented by dotted lines).

Strengths and Limitations

This analysis is an alternative way to study the natural history of LOVA. The statistical methods used in this paper reflect the diverse categorical and continuous data. The main strength of this method is its lack of bias, being a data driven cluster analysis.

Some clinical and radiological features were not included in the cluster analysis due to low occurrence and due to statistical restrictions on the number of variables used. The analysis of

a patients referred to a single centre is a limitation of the study, in addition to the low numbers in some subgroups.

Future analysis would involve validating this cluster model prospectively, both clinically and against alternative classifications such as primarily radiological ones.

Conclusion

Patients with ventriculomegaly commonly have a post-natal event (e.g after premature birth), followed by four main typical presentations that span different age groups including (1) incidental ventriculomegaly with or without headache, (2) highly symptomatic which can lead reduced consciousness and coma if untreated (3) early-presenting with milder symptoms with of headache and nausea and (4) late-presenting with features common to NPH. The different symptomatic subgroups are likely to represent the various degrees (and time of presentation) of decompensation in chronic hydrocephalus.

We find that ICPM is useful to determine sub-types and may be a useful guide for management. Some groups can be managed conservatively, whilst others benefit from ETV or VP shunting. The surgical interventions may not be equally effective across the sub-groups, presenting an avenue for further research. The identified sub-types provide further insight into the natural history of chronic ventriculomegaly in adults.

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