CSF SHUNTING FOR PSEUDOTUMOR CEREBRI SYNDROME  
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I. HISTORY  
Ventriculo-cisternostomy was first introduced by Arne Torkildsen in 1937 [1]. This technique, consisting of a simple shunt tube without valves, was an advance over previous methods because cerebrospinal fluid (CSF) was shunted to and from spaces that normally contained CSF. In the 1950's, development of functional one-way valves led to the introduction of ventriculo-venous shunting techniques [2]. All shunts are pressure-regulated and generally employ slit, spring-ball or diaphragm valves. Lumboperitoneal shunting (LPS) has the advantage over ventricular procedures of minimizing intracranial complications and has also been shown to have a lower incidence of infection and malfunction [2, 3, 4]. LPS, however, is not appropriate for the treatment of non-communicating hydrocephalus and also carries a higher incidence of complications in children than in adults [3, 4]. Possible indications for LPS include communicating hydrocephalus, CSF fistula and pseudotumor cerebri syndrome (PTC).

II. CSF SHUNTING FOR PTC  
There are few reports analyzing the use of CSF shunting procedures for the treatment of pseudotumor cerebri syndrome (PTC) [5, 6, 7, 8]. All are retrospective and non-randomized. These studies are sometimes difficult to compare or contrast because of differences among them:

A. Different techniques
   - Johnston: percutaneous LP, valved LPS, cisternal atrial, VP and VA
   - Rosenberg: 73 LPS, 8 VP, 1 VA, 1 VJ
   - Eggenberger: all LPS
   - Burgett: all LPS

B. Different patient groups
   - idiopathic PTC: Rosenberg  
   - all causes: Johnston, Eggenberger, Burgett

C. Different indications
   - Johnston: papilledema, visual symptoms and/or loss, concern regarding steroids
   - Rosenberg: headache in 18/27 (67%), visual loss in 14/27 (52%)
   - Burgett: headache in 25%, visual loss in 70%

D. Proportion of patients treated with shunt
   - Johnston: 36/41 (88%)
   - Burgett: 30/168 (18%)
   - others: not available

E. Data concerning optic nerve function
   - Johnston: not available
   - Rosenberg: "improved, same or worse" without details
   - Eggenberger: VA, VF and color combined
   - Burgett: detailed data regarding pre-op and post-op VA and VF

III. REVIEW OF DATA FROM OUR SERIES [8]

A. Methods
   We reviewed the charts of 168 patients with PTC seen by 1 author (VP) between 1983-1995. Of these, 30 patients treated with LPS were identified. Seventeen of the 30 had full neuro-eye evaluation pre and post-op. We recorded: age, sex, race, etiology of PTC, indications for LPS, effect on symptoms and on optic nerve function and appearance, number of revisions with indications and duration of follow-up.

B. Results
   Mean age was 32.9 years with a range of 10-68 years. Twenty-eight of 30 patients were female. Etiologies were varied: obesity (23), COPD (2), steroid withdrawal (1), menarche (1), and idiopathic (3).

C. Effect on symptoms (N=17)
   - There was significant improvement of symptoms in 82% of patients. One patient had persistent high pressure headache and one suffered a low pressure syndrome.

D. Effect on vision (N=34 eyes)
   1. visual acuity
      - All 20 eyes with 20/25 were stable post-operatively of eyes with 20/30, 10/14 (71%) were improved. Of these, 4/14 improved by 7 lines. One eye was worse (in patient who had already undergone optic nerve sheath fenestration).
   2. visual field (Goldmann)
      - Of 26 eyes with an abnormal pre-operative visual field: 5 (19%) normalized, 13 (50%) were significantly improved, 8 (30%) were unchanged and none worsened.
   3. papilledema
      - There was complete or near-complete resolution in 24/25 eyes (96%) with severe papilledema and complete resolution in 7 eyes with mild or moderate edema.

E. Revisions (N=126)
   1. indications (when available)
      - recurrent (ICP (13) (ICP (2) CSF fistula (1) sciatica (1) tonsillar herniation (0))
   2. rate
      - mean: 4.2 shunt procedures/patient median: 1/patient mean with 4 outliers removed: 2.5/patient (86/126 revisions were in 4 patients)
   3. interval
      - average revision time: 18 months
IV. COMPARISON OF OUR DATA WITH OTHER SERIES

A. Efficacy

Johnston: "rapid and complete relief" in 36/36 (100%)
Rosenberg: visual improvement in 13/37, worse in 6/37, no change in 18/37
Eggenberger: relief of h/a in 18/18 (100%) visual improvement in 16/28 eyes (68%)

B. Revisions

Rosenberg
2.2 revisions/patient
average time: 9 months
no revisions in 18/37 (49%);
1 or 2 in 13 (35%)
> 2 revisions in 6/37 (17%)
for shunt failure in 31 (55%)

Eggenberger
2.4 revisions/patient
average (median) time 11 months
no revisions in 12/27 (44%);
1 or 2 in 8 (30%)
> 2 revisions in 7/27 (26%)
for shunt failure in 43/66 (65%)

Burgett
4.2 revisions/patient (2.5 without 4 outliers)
average time 18 months
no revisions in 11/30 (37%); 1 or 2 in 6 (20%)
> 2 revisions in 13/30 (43%)

3 series combined (N=94 patients)
2.9 revisions/patient
average time 12 months
no revisions in 44%
> 2 revisions in 28%
shunt failure was the indication in 53%

C. Serious complications (shunt infection)

Johnston: none
Rosenberg: 3.6%
Eggenberger: 1.5%
Burgett: 1%

V. PREDICTORS OF SHUNT FAILURE

There is clearly a subgroup of patients who seem to tolerate shunts poorly. Ideally we would like to be able to identify these patients before embarking on a first shunting procedure but the factors that contribute to the need for frequent revisions are unclear. The data of Eggenberger et al suggested that younger age may be a factor although this was not borne out in other studies. These authors suggested using the duration of the initial shunt for this purpose. In their series, patients who required shunt revisions typically did so within the first 2-3 months following the first procedure and always within the first year. Our data did not support this finding. We suggest that the need for >3 revisions may be a better predictor of the need for multiple additional procedures.

VI. COMPLICATIONS OF LPS

The most common complications of LP shunting are obstruction and intracranial hypotension. Numerous other complications have been reported, including lumbar radiculopathy, shunt-related abdominal pain, shunt or disc space infection, migration or dislocation of the peritoneal end of the catheter, and one case of rupture of an intracranial aneurysm [9, 10, 11, 12, 13, 14]. Tonsillar descent secondary to LPS has also been well documented. Initially considered a rare complication, this form of acquired Chiari I malformation was found in approximately half of a group of 143 pediatric patients, most of whom were shunted for hydrocephalus [15]. In another series, 7 of 10 pediatric patients developed this complication [16]. In most patients this is an asymptomatic finding. Some patients require treatment [16, 17] and one child with hydrocephalus and Crouzon's disease died as a result of such an acquired malformation [15]. This complication does not appear to be a problem in patients shunted for PTC.

A few reports in the literature suggest acute visual loss as a complication of shunting [18, 19]. These cases represent shunt failures, with visual loss secondary to recurrent PTC, rather than a true complication of LPS. In some reported cases, visual loss was progressive despite a shunt that was believed to be functioning [20].

VII. SUMMARY

LPS is generally a safe and effective procedure for treating the symptoms and visual loss associated with PTC. The incidence of potentially serious complications (mostly meningitis) is low (1.5%). The main drawback of LPS for PTC is the need for multiple shunt revisions in some patients. The most common indication for shunt revision is malfunction (ie recurrent ICP). Approximately 50% of PTC patients are satisfactorily treated with just one shunting procedure. Three or more shunt revisions are needed in 28%, including a smaller subgroup requiring multiple, frequent revisions. The factors that contribute to poor shunt tolerance are unclear. Predictors of the need for multiple shunt revisions may include younger age, short interval from initial to 2nd shunt and > 2 revisions.

VIII. FUTURE DEVELOPMENTS

Advances in techniques for stereotactic guidance have been applied to the placement of VP shunts. This procedure may be associated with a lower failure rate than lumbar shunts in the treatment of patients with PTC. Tulipan et al reported resolution of headache in 6 of 7 PTC patients treated with stereotactic VP shunting [21]. The recent development of an externally programmable valve should reduce the need for revisions to treat low pressure syndromes. The combination of these 2 techniques may prove a more satisfactory long-term treatment for patients with intractable PTC.
References