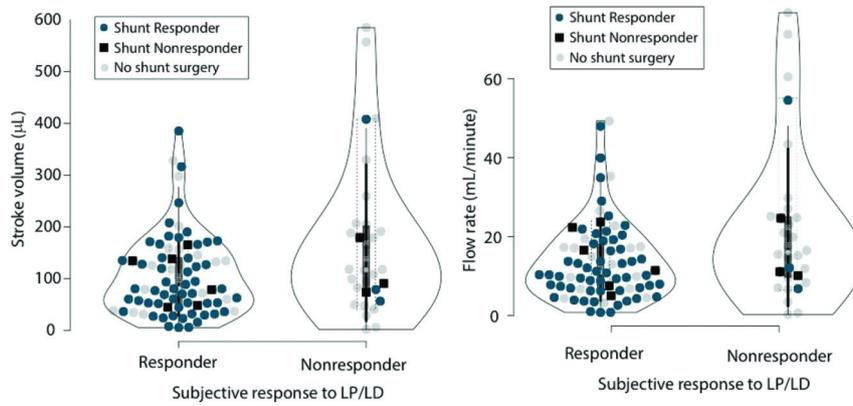
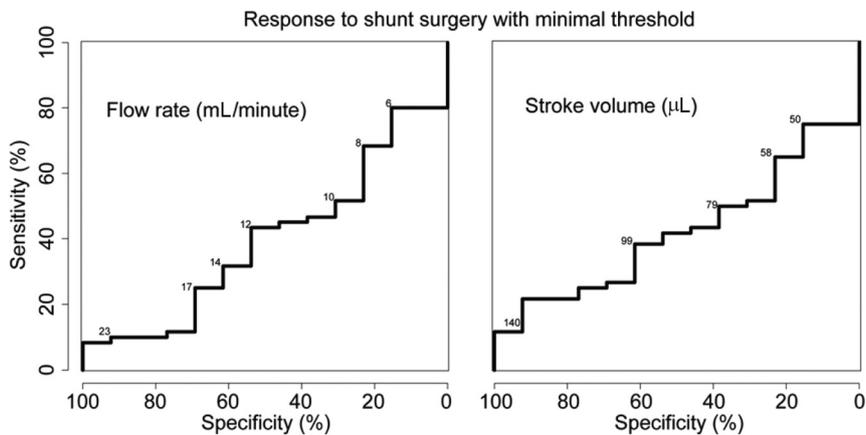


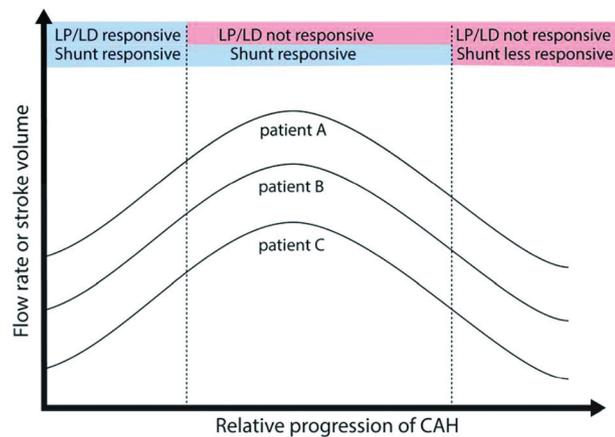
ON-LINE FIG 1. The aqueductal CSF flow and shunt response measured by subjective improvement and the TUG score improvement. A, The distributions of aqueductal stroke volume (*upper*) and flow rate (*lower*) are compared between shunt responders and nonresponders. The aqueductal flow rate does not show a significant difference between shunt responders and nonresponders. (Wilcoxon rank sum test, $P = .38$ for stroke volume and $.23$ for flow rate). B, The aqueductal stroke volume (*upper*) and flow rate (*lower*) are plotted against the quantitative improvement of the TUG score (*y-axis*) after shunt surgery. The Pearson correlation between the TUG score improvement and the stroke volume and flow rate are -0.191 and -0.198 , respectively. C, Twenty percent or greater improvement in the TUG score after shunt surgery was used to define shunt responders, and $<20\%$ improvement in the TUG score after shunt surgery was used to define shunt nonresponders. The 2 groups exhibit an overlapping distribution of aqueductal flow rate and stroke volumes. D, "Tinetti score deficit" is defined as the difference between a perfect Tinetti score and the patient's Tinetti score. Twenty percent or greater improvement in the Tinetti score deficit after shunt surgery was used to define shunt responders, and $<20\%$ improvement in the Tinetti score deficit after shunt surgery was used to define shunt nonresponders. Again, the 2 groups exhibit an overlapping distribution of the aqueductal flow rates and stroke volumes.



ON-LINE FIG 2. Shunt responses among LP/LD responders and nonresponders. The LP/LD responders have a significantly higher chance of responding to shunt surgery than LP/LD nonresponders (Fisher exact test, $P = .03063$). Regardless of the LP/LD results, all the patients with flow rates of >25 mL per minute or stroke volume of >180 μ L have favorable shunt surgery outcomes.



ON-LINE FIG 3. The receiver operating characteristic analysis using varying aqueductal flow rates and stroke volume thresholds, denoted as numbers on the curves, in determining shunt responders and nonresponders.



ON-LINE FIG 4. Diagram of aqueductal flow over the disease course of CAH. In this diagram, patients with CAH exhibit increasing aqueductal flow during the early disease course, but after a certain point, the aqueductal flow starts to decrease. There is significant heterogeneity in baseline aqueductal flow among individuals, which makes it difficult to determine the extent of disease progression from single snapshots of flow characteristics. LP and LD have high positive predictive values but low negative predictive values because they miss patients who can benefit from shunt surgery.

On-line Table 1: Flow rate^a

	Top 25%	Midhigh 25%	Midlow 25%	Bottom 25%	Normal at Baseline
Baseline Tinetti Test score	18.15 ± 2.43 (49)	15.59 ± 3.02 (29)	14.18 ± 1.39 (32)	12.89 ± 1.36 (35)	NA
Baseline TUG score	17.7 ± 2.55 (38)	16.71 ± 2.62 (35)	14.88 ± 2.28 (35)	12.81 ± 1.25 (36)	NA
Baseline MMSE score	16.79 ± 1.84 (54)	15.12 ± 2.87 (24)	15.07 ± 2.22 (35)	13.03 ± 1.86 (33)	NA
ΔTUG post-LP/LD	13.64 ± 1.35 (29)	16.09 ± 2.49 (29)	16.76 ± 2.66 (28)	18.29 ± 3.36 (29)	NA
ΔTUG postshunt	11.49 ± 1.82 (16)	11.26 ± 1.53 (16)	16.94 ± 3.4 (17)	15.34 ± 3.24 (15)	NA
ΔTinetti post-LP/LD ^b	16.61 ± 2.39 (26)	11.41 ± 1.32 (25)	18.45 ± 2.42 (44)	11.82 ± 2.91 (7)	16.18 ± 3.95 (9)
ΔTinetti postshunt ^b	15.15 ± 3.71 (15)	11.17 ± 2.18 (13)	13.06 ± 2.05 (23)	15.30 ± 3.88 (5)	18.03 ± 4.74 (6)
ΔMMSE postshunt ^c	11.47 ± 2.25 (8)	15.15 ± 2.10 (11)	6.29 ± 1.42 (3)	14.12 ± 4.42 (8)	11.79 ± 3.61 (4)

Note:—NA indicates not applicable.

^a Data are means in milliliters/minute (No. of patients). Average aqueductal flow rate of each quartile for baseline Tinetti, TUG, and MMSE scores (upper 3 rows) and improvement in TUG, Tinetti, and MMSE scores on LP/LD and shunt surgery (lower 5 rows). Patients are divided into 4 groups based on the Tinetti, TUG, and MMSE scores at imaging and the Tinetti, TUG, and MMSE changes (ΔTinetti, ΔTUG, and ΔMMSE) after LP/LD and shunt surgery. A quartile with higher baseline scores (upper 3 rows) or greater improvement (lower 5 rows) after LP/LD or shunt surgery is defined as "Top 25%." To prevent bias from the ceiling effect of ΔTinetti and ΔMMSE, we processed patients having perfect scores both at pre- and post-LP/LD or shunt surgery separately in the "Normal" column.

^b ΔTinetti is defined as $(\text{Tinetti}_{\text{post-procedure}} - \text{Tinetti}_{\text{pre-procedure}}) / \text{Tinetti}_{\text{pre-procedure}}$; Tinetti score changes of 1 or 0 were substituted with ΔTinetti = 0.

^c ΔMMSE is defined as $(\text{MMSE}_{\text{post-procedure}} - \text{MMSE}_{\text{pre-procedure}}) / \text{MMSE}_{\text{pre-procedure}}$.

On-line Table 2: Stroke volume^a

	Top 25%	Midhigh 25%	Midlow 25%	Bottom 25%	Normal at Baseline
Baseline Tinetti Test score	142 ± 19 (49)	116 ± 23 (29)	103 ± 11 (32)	96 ± 11 (35)	NA
Baseline TUG score	135 ± 20 (38)	126 ± 20 (35)	113 ± 17 (35)	96 ± 10 (36)	NA
Baseline MMSE score	125 ± 14 (54)	117 ± 24 (24)	113 ± 17 (35)	99 ± 14 (33)	NA
ΔTUG post-LP/LD	102 ± 11 (29)	124 ± 19 (29)	130 ± 23 (28)	134 ± 23 (29)	NA
ΔTUG postshunt	83 ± 14 (16)	89 ± 13 (16)	125 ± 28 (17)	114 ± 21 (15)	NA
ΔTinetti post-LP/LD ^b	124 ± 16 (26)	87 ± 13 (25)	137 ± 18 (44)	85 ± 21 (7)	136 ± 37 (9)
ΔTinetti postshunt ^b	108 ± 27 (15)	81 ± 15 (13)	97 ± 14 (23)	116 ± 30 (5)	155 ± 49 (6)
ΔMMSE postshunt ^c	88 ± 20 (8)	110 ± 17 (11)	119 ± 42 (3)	93 ± 32 (8)	93 ± 32 (4)

Note:—NA indicates not applicable.

^a Data are means in milliliters/minute (No. of patients). Average aqueductal stroke volume of each quartile for baseline Tinetti, TUG, and MMSE scores (upper 3 rows) and improvement in TUG, Tinetti, and MMSE scores on LP/LD and shunt surgery (lower 5 rows). Patients are divided into 4 groups based on the Tinetti, TUG, and MMSE scores at imaging and the Tinetti, TUG, and MMSE changes (ΔTinetti, ΔTUG, and ΔMMSE) after LP/LD and shunt surgery. A quartile with higher baseline scores (upper 3 rows) or greater improvement (lower 5 rows) after LP/LD or shunt surgery is defined as "Top 25%." To prevent bias from the ceiling effect of ΔTinetti and ΔMMSE, we processed patients having perfect scores both at pre- and post-LP/LD or shunt surgery separately in the "Normal" column.

^b ΔTinetti is defined as $(\text{Tinetti}_{\text{post-procedure}} - \text{Tinetti}_{\text{pre-procedure}}) / \text{Tinetti}_{\text{pre-procedure}}$; Tinetti score changes of 1 or 0 were substituted with ΔTinetti = 0.

^c ΔMMSE is defined as $(\text{MMSE}_{\text{post-procedure}} - \text{MMSE}_{\text{pre-procedure}}) / \text{MMSE}_{\text{pre-procedure}}$.