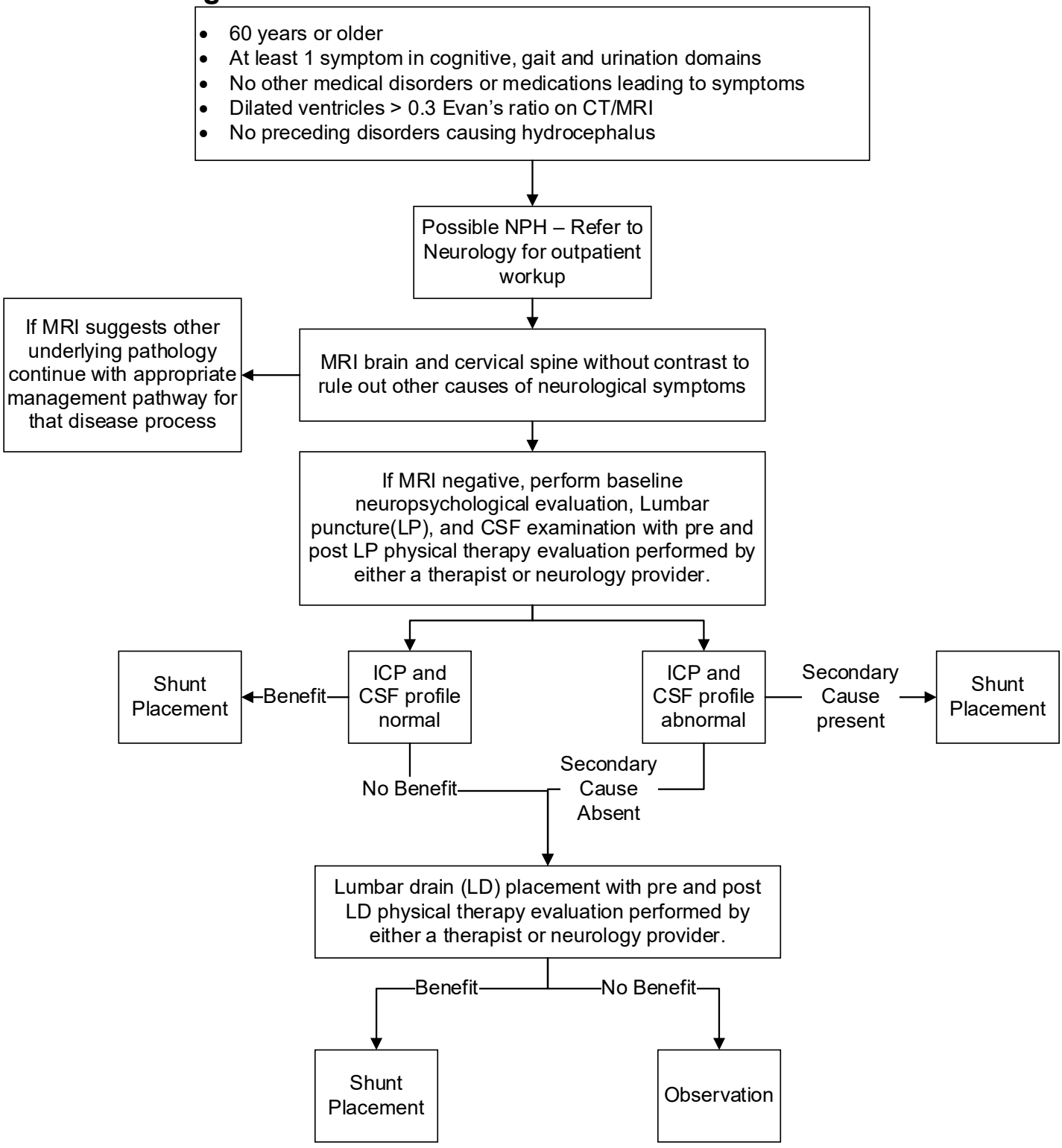
Clinical Pathways Program

Guideline: Normal Pressure Hydrocephalus, Adult

Updated: March 19, 2021

Clinical algorithm:



Clinical guideline summary

CLINICAL GUIDELINE NAME: Normal Pressure Hydrocephalus Guideline

PATIENT POPULATION AND DIAGNOSIS: Adults >18 year of age, idiopathic normal pressure hydrocephalus (iNPH) -For information on diagnosis and symptoms, please see DiagnosisandSymptoms

APPLICABLE TO: All Spectrum Health Sites

BRIEF DESCRIPTION: To present recommendations for diagnosis and treatment of idiopathic normal pressure hydrocephalus (iNPH) and determine predictors of shunting effectiveness, based on a systematic review and analysis of the evidence.

Definitions

iNPH: Idiopathic Normal Pressure Hydrocephalus
CT: Computed Tomography
MRI: Magnetic Resonance Imaging
CSF: Cerebrospinal Fluid
Ro : Outflow Resistance

OVERSIGHT TEAM LEADER(S): Rushna Ali, MD, Aiesha Ahmed, MD, MBA

OWNING EXPERT IMPROVEMENT TEAM (EIT): N/A

MANAGING CLINICAL PRACTICE COUNCIL (CPC): Neurosciences CPC

CPC APPROVAL DATE:3/19/2021

OTHER TEAM(S) IMPACTED (FOR EXAMPLE: CPCs, ANESTHESIA, NURSING, RADIOLOGY): Interventional Radiology, Neurology

IMPLEMENTATION DATE: January 2021

LAST REVISED: 3/19/2021

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Clinical pathways clinical approach

TREATMENT AND MANAGEMENT:

Diagnosis and Symptoms:
Idiopathic Normal Pressure Hydrocephalus (iNPH) is a clinical syndrome that includes dementia and urinary incontinence in addition to gait disturbance as major manifestations in the absence of preceding disorders, including subarachnoid hemorrhage and meningitis, but with ventricular dilation caused by an impairment of CSF circulation. iNPH develops in elderly patients, and its symptoms usually progress slowly. The symptoms can be improved by appropriate CSF shunt surgery.

Symptoms: Idiopathic Normal pressure hydrocephalus (iNPH) comprises of the clinical triad of gait impairment, urinary incontinence and cognitive impairment. Gait disturbance and cognitive impairment are the most prevalent. Apraxic/ataxic gait with a small-stepped, magnetic, and broad-based nature is characteristic. Freezing is apparent on the initiation of walking, walking in a narrow space, and turning around. Cognitive impairment is demonstrable on neuropsychological tests. Urinary incontinence features include overactive bladder, mainly manifesting as increased nocturnal urinary frequency and urge urinary incontinence. The following scale has been used by groups to determine a severity score.

Gait		Urinary incontinence		Cognition	
Normal	5	None	5	None	6
Walk with support	4	Rare incontinence	4	Subjective decrease in memory	5
Requires cane	3	Occasional incontinence	3	Objective decrease in memory but independent	4
Support by person	2	Constant incontinence	2	Partial loss of independence	3
Wheelchair or bed	1	Permanent catheter	1	Disoriented x 2	2
				Institutionalized secondary to dementia	1

Other causes of these symptoms should be ruled out. Significant number of patients with suspected NPH will harbor alternative diagnoses. Identification of contributing/confounding conditions will support the meticulous work-up necessary to appropriately manage patients without NPH while optimizing clinical responses to shunting in correctly diagnosed patients.

Radiological features: MRI is recommended for diagnosis. CT may be used, when MRI is contraindicated. The diagnosis of iNPH should be suspected based on both symptomatic and imaging findings. It should not be diagnosed based only on imaging findings, if there are no characteristic symptoms. Evans index (ratio of the maximum width of the frontal horns to the maximum width of the inner table of the cranium) of greater than 0.3 is a hallmark of hydrocephalus. MRI brain and cervical spine should also be used to evaluate for other causes leading to these symptoms including lesions such as stroke, tumors, obstructive hydrocephalus or cervical stenosis.

Tap Test and Lumbar Drain Test: A tap test is recommended to confirm the diagnosis and determine candidacy for shunt placement. This should be performed on an outpatient basis. A high-volume lumbar puncture is performed where 30-50 cc of CSF is removed. Baseline Neuropsychological evaluation is obtained to assess for underlying cognitive disorders which may or may not be related to NPH. Formal physical therapy testing is performed before and after the tap test and lumbar drain trial to determine response and benefit. This can be performed by a provider or physical therapist.

When a tap test is negative, there are three options: a repeat tap test; a lumbar drain trial; and reconsider the diagnosis and observe further. When repeating a tap test, use a thicker needle and remove a greater amount of CSF than during the first test. A lumbar drain trial removes 10 cc/hr for 48-72 hours. The patient obtains a lumbar drain placed by Interventional Radiology and is admitted to the ICU for monitoring and lumbar drainage. Complications such as disconnection or fracture of the indwelling catheter, radicular pain, or meningitis have been reported in 2–8% of patients. In addition to the clinical response to CSF removal, studies have investigated CSF dynamics. Ro is a measure of outflow resistance during CSF drainage. Elevated Ro has a positive predictive value that could be as high as 94%, but negative predictive value does not exceed 19%. In patients with suspected iNPH, those with elevated Ro are probably more likely to respond to shunting than those without elevated Ro, but lower Ro does not preclude shunt responsiveness. In a population preselected on the basis of clinical and imaging criteria; positive predictive value of the tap test has been reported to be 88% whereas negative predictive value only 18%, resulting in an overall accuracy of 53%. Therefore, patients who improve after tap tests may be more likely to respond to shunting, but negative tap tests do not preclude a response to shunting.

It is important to remember that a complete and thorough NPH work up should be performed on an outpatient basis since an acute illness while inpatient can affect the result of the testing significantly.

Treatment

Shunt surgery: Cerebrospinal fluid diversion can be performed using the following types of shunts; Ventriculo-peritoneal, ventriculo-atrial, ventriculo-pleural and lumbo-peritoneal shunt procedures. Attention must be paid to status of caregiving and consent of the family. Physical therapy and rehabilitation should be performed before and after surgery, whenever necessary. Evaluation of the effects should be followed until one year after surgery and on an as-needed basis thereafter using formal physical therapy and neuropsychological testing.

Studies have reported clinical improvement of iNPH after shunting, but none to date has been designed to provide high-level evidence of efficacy. It should be recognized that the use of ventricular shunting for iNPH is based largely on uncontrolled observational studies of clinical response. Published evidence indicates that there is a decrease of response to shunting after 6 months and fewer than half of patients were considered to be improved in all presenting iNPH symptoms after 18 months.

Predictors of successful outcome: There is evidence that improvement in gait following the CSF tap test or external lumbar drainage may predict a good response to shunting. MRI-measured high CSF velocity through the aqueduct and abnormal intracranial CSF hydrodynamics are also predictive of good response to shunting.

In patients with suspected iNPH, there is insufficient evidence to determine whether advanced age and presence of 3 or more major medical comorbidities are likely to respond less favorably to shunting than those with fewer comorbidities.

Therefore, in conclusion, shunting is possibly effective for patients with iNPH with 96% chance of subjective improvement and 83% chance of improvement on the timed walk test at 6 months in appropriately selected patients. There is approximately an 11% risk of serious adverse events which include meningitis, ventriculitis, shunt failure and hardware infections which should be considered when surgical recommendations are made.

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