CASE RECORDS of the MASSACHUSETTS GENERAL HOSPITAL

Case 40-2016: A 14-Month-Old Girl with Recurrent Vomiting

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PRESENTATION OF CASE

Dr. Alessio Morley-Fletcher (Pediatrics): A girl 14 months 19 days of age was admitted to a community hospital affiliated with this hospital because of episodes of recurrent vomiting.

The patient had been well until she was 11 months 12 days of age, when intermittent vomiting developed. On the fourth day of symptoms, she was seen by a pediatrician. The stools were reportedly soft but formed, with no diarrhea. A diagnosis of viral gastroenteritis was made, and dietary changes were advised. The patient returned home, and the symptoms resolved. On routine examination when she was 12 months of age, her parents reported that she could perform activities that were developmentally appropriate for her age, including walking, holding on, and using a cup; physical examination was normal. Routine childhood immunizations were administered.

During the next 2 months, the patient had two episodes of recurrent vomiting; the episodes lasted 3 days and 5 days and were associated with lethargy and decreased oral intake, as well as with diarrhea on some but not all occasions. During each episode, an examination performed in the emergency department of the other hospital was normal, and no fever or abdominal pain was present. Laboratory test results are shown in Table 1. A diagnosis of acute gastroenteritis with dehydration was made during each episode. Ondansetron and intravenous fluids were administered, and the patient returned home. The episodes resolved spontaneously. Three weeks before the current admission, milk formula was stopped and a soy-based formula was begun.

Eighteen days before the current admission to the other hospital, the patient was seen by a pediatric gastroenterologist at this hospital. Examination was normal, and a possible diagnosis of vomiting caused by nonketotic hypoglycemia was made. Her parents were advised to seek a medical evaluation for her, including metabolic studies, if another episode occurred.

On the day of admission, four episodes of nonbloody, nonbilious vomiting occurred in the evening, and the patient was taken to the emergency department of the other hospital. The patient had been born at full-term gestation, without complications during gestation or delivery. The weight at birth was 3.3 kg, and the length 50.2 cm. The Apgar scores at 1 minute and 5 minutes were 8 and 9, respec-

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Table 1. Laboratory Data Obtained at the Other Hospital.*

Variable	Reference Range, Age-Adjusted†	2nd Episode, 2 Mo before Admission	3rd Episode, 1 Mo before Admission	4th Episode, Day of Admission	16 Hr after Presentation
Age		12 mo 27 days	13 mo 23 days	14 mo 19 days	
Platelet count (per mm^3)	135,000–400,000				476,000
Sodium (mmol/liter)	136–145	138	138	144	141
Potassium (mmol/liter)	3.5–5.2	4.6	4.4	5.1	4.5
Chloride (mmol/liter)	99–109	96	99	101	102
Carbon dioxide (mmol/liter)	20–31	16	24	23	23
Plasma anion gap	4–14	26	15	20	15
Urea nitrogen (mg/dl)	9–23	14	9	15	5
Creatinine (mg/dl)	0.5–1.3	0.35	0.29	0.29	0.20
Glucose (mg/dl)	74–106	69	84	100	81
Calcium (mg/dl)	8.7–10.4	10.8	10.8	11.4	10.7
Protein (g/dl)					
Total	5.9–7.0			7.3	
Albumin	3.4–4.8			5.3	
Lactate (mmol/liter)	0.5–2.5			5.0	2.9
Alkaline phosphatase (U/liter)	70–250			264	
Total carnitine (nmol/ml)	35–84			27	
Free carnitine (nmol/ml)	24–63			18	
Acylcarnitine (nmol/ml)	4–28			9	
Acylcarnitine:free carnitine ratio	0.1–0.8			0.5	
Pyruvate (mg/dl)	0.7–1.4				2.9
Serum β-hydroxybutyrate (mmol/liter)	<0.4				0.8
Venous blood gases					
Inhaled oxygen (%)					Not specified
pH	7.35–7.45				7.43
Partial pressure of carbon dioxide (mm Hg)	38–52				33
Partial pressure of oxygen (mm Hg)	42–54				41
Oxygen saturation (%)					76.6

* To convert the values for urea nitrogen to millimoles per liter, multiply by 0.357. To convert the values for creatinine to micromoles per liter, multiply by 88.4. To convert the values for glucose to millimoles per liter, multiply by 0.05551. To convert the values for calcium to millimoles per liter, multiply by 0.250. To convert the values for lactate to milligrams per deciliter, divide by 0.1110. To convert the values for pyruvate to micromoles per liter, multiply by 113.6.

† Reference values are affected by many variables, including the patient population and the laboratory methods used. The ranges shown are from the other hospital and are age-adjusted for patients who are not pregnant and do not have medical conditions that could affect the results. They may therefore not be appropriate for all patients.

tively. Jaundice was noted during the newborn period. The patient was breast-fed exclusively for 8 weeks and received supplemental formula thereafter, and solid foods were introduced at 5 months. She had mild facial eczema. She received no medications and had no known allergies. Her father had gallstones and had had pancreatitis in his 20s, her paternal grandmother had gallstones, her paternal grandfather had colon cancer, both grandfathers had diabetes mellitus, her

maternal grandfather had arthritis, and a distant paternal cousin had Crohn's disease; there was no family history of celiac disease.

On examination in the emergency department, the patient vomited and had dry heaves intermittently. There was a regular heart rate and rhythm, and the temperature, respiratory rate, and remainder of the general examination were normal. Blood levels of amylase, lipase, ammonia, alanine aminotransferase, aspartate aminotransferase, total bilirubin, direct bilirubin, pyruvate kinase, and amino acids were normal; other test results are shown in Table 1. Urinalysis revealed 1+ ketones and leukocyte esterase by dipstick and 5 white cells per high-power field; screening for toxins in the urine was negative, and a urine culture was sterile. A presumptive diagnosis of gastroenteritis was made, and the patient was admitted to the pediatric inpatient unit for observation and rehydration.

On admission to the pediatric inpatient unit, additional history was obtained from the patient's parents. They reported that in retrospect, during the previous 3 months, the patient had stopped pulling to stand or attempting to walk and was only crawling (and when doing so appeared to be unsteady, with occasional "face plants"). They noted occasional "shakes" when she woke up in the morning, which consisted of symmetric movement for a few seconds that would recur over a 10-minute period, without loss of tone or consciousness.

On examination, the patient was alert and interactive. Frontal bossing was present and the fontanelle was full, with mild pulsations that were visible anteriorly when she was in the supine position. The weight was 9.9 kg, the length 78.3 cm, and the head circumference 51.5 cm (>95th percentile). The eye movements were normal, the pupils were equally reactive, and there was no horizontal or vertical limitation or nystagmus. The legs had decreased muscle tone, with brisk 3+ patellar reflexes, and the plantar reflexes were flexor. There was no crossed adductor reflex (i.e., no contraction of both hip adductors when the knee jerk was elicited on either side, which is a sign of pyramidal tract dysfunction in patients older than 8 months of age) and no ankle clonus. The remainder of the examination was normal, including evaluation of the muscle tone in the shoulder girdle. The hematocrit, hemoglobin level, red-cell indexes, white-cell count, and white-cell differential count were normal; additional laboratory test results are shown in Table 1. The patient's pediatrician was contacted for additional information about her growth (Fig. 1).

A diagnostic test was performed, and the patient was transferred to this hospital for further evaluation and treatment.

DIFFERENTIAL DIAGNOSIS

Dr. Jess L. Kaplan: This patient is a 14-month-old girl with episodes of recurrent vomiting. Her current presentation is the fourth such episode to occur during a period of 3 months; the previous episodes of vomiting resolved with supportive care, fluid resuscitation, and antiemetic medication. Recurrent vomiting is not an uncommon symptom in infants and children and is a relatively common reason for referral to a pediatric gastroenterologist.

RECURRENT VOMITING IN CHILDREN

The differential diagnosis of recurrent vomiting during early childhood is broad and includes gastrointestinal, metabolic, and neurologic causes, among others. Temporal patterns can be helpful in determining cause. A chronic pattern of recurrent vomiting often has a gastrointestinal cause. A cyclical pattern of recurrent vomiting is more likely to be due to a nongastrointestinal condition.[1] The pattern in this case appears to be more cyclical than chronic. Key clinical features help to identify serious causes for which the delay of diagnosis may have clinical consequences (Table 2). For this patient, I believe the key clinical features include the rapid increase in head circumference, developmental regression, and certain abnormal findings on laboratory tests and physical examination.

GASTROINTESTINAL CAUSES

Recurrent gastrointestinal infection (more likely viral infection than bacterial or parasitic infection) was the suspected cause of this patient's first three episodes of vomiting. The presence of dehydration, which is indicated in this patient by the low serum carbon dioxide level and high plasma anion gap at the time of the second presentation, is common in but not specific for

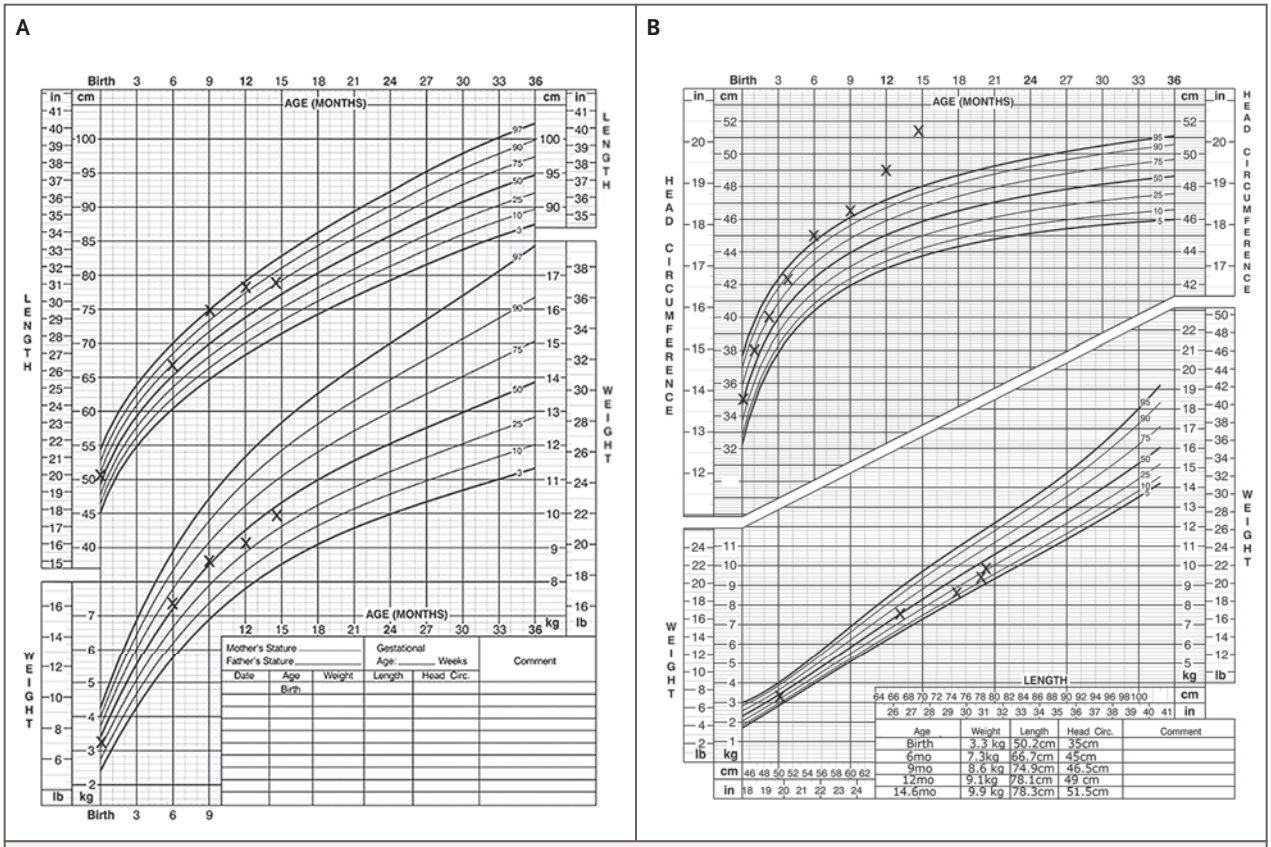


Figure 1. Growth Charts.

Panel A shows the patient's charts for length and weight in relation to age. Since birth, the weight has remained near the 50th percentile for her age and the length has ranged between the 50th percentile and greater than the 95th percentile, with the most recent measurement, obtained when she was 14 months of age, falling in the 75th percentile. Panel B shows the patient's chart for head circumference in relation to age and for weight in relation to length. The head circumference was in the 50th percentile at birth but was in the 95th percentile when she was 9 months of age, with continued increase in percentile between 9 and 14 months of age. Charts containing the percentiles were developed by the National Center for Health Statistics in collaboration with the National Center for Chronic Disease Prevention and Health Promotion (www.cdc.gov/growthcharts).

gastrointestinal infection. However, many features of this patient's clinical presentation do not support a diagnosis of recurrent infection. Viral gastroenteritis is frequently accompanied by abdominal cramping and diarrhea, features that were not consistently present in this patient. The absence of fever and of known sick contacts also makes recurrent gastrointestinal infection unlikely. Although gastrointestinal infection is common and highly contagious in young children and could reasonably explain one or two of the episodes, it is unlikely that this child would have four distinct episodes of vomiting due to one or more gastrointestinal infections over a span of 3 months. Finally, important clinical features, such as developmental regression and worsening macrocephaly, cannot be explained by recurrent gastrointestinal infection alone. Young children with nongastrointestinal infections, such as pneumonia, acute otitis media, urinary tract infection, and meningitis, also frequently present with vomiting and without other focal symptoms. However, these conditions are often accompanied by fever and leukocytosis and are supported by findings on physical examination, laboratory tests, or both, and these features were not present in this patient.

Common noninfectious gastrointestinal causes of recurrent vomiting in infants and toddlers include gastroesophageal reflux and mucosal

Table 2. Key Signs and Symptoms in Children with Vomiting and Associated Serious Conditions.

Sign or Symptom	Associated Serious Condition
Bilious vomiting	Bowel obstruction
Abdominal distention	Bowel obstruction and peritonitis
Hematemesis	Mucosal injury of the upper gastrointestinal tract, esophageal varices, and prolapsing gastropathy
Hematochezia	Inflammatory bowel disease, gastrointestinal infection, intussusception, and midgut volvulus
Vomiting in the night or early morning	Increased intracranial pressure
Vomiting without preceding nausea (in older children) and positional triggers	Increased intracranial pressure
Associated headache (in older children)	Increased intracranial pressure
Lethargy and hypotension	Metabolic disorder, adrenal crisis, and systemic illness
Focal neurologic symptoms and developmental delay or regression	Metabolic disorder, neurologic conditions involving increased intracranial pressure, and toxic ingestion
Weight loss	Systemic illness or metabolic disorder

inflammation of the upper gastrointestinal tract due to peptic injury or allergic disease, such as eosinophilic gastroenteritis or eosinophilic esophagitis. Gastroesophageal reflux typically improves gradually during the first year of life and rarely initially occurs in patients who are 11 months of age.[2] Vomiting caused by gastroesophageal reflux and eosinophilic esophagitis can be severe, but the vomiting is not typically episodic or as severe as it was in this patient and rarely leads to dehydration and hospitalization. Patients with the food protein–induced enterocolitis syndrome (FPIES) present with profuse episodic vomiting, lethargy, and dehydration after the ingestion of the offending food antigen[3]; FPIES is a possible cause in this case. However, presentation at 11 months of age is relatively late, and the absence of a consistent and specific food trigger for the vomiting makes FPIES unlikely in this patient. Episodic vomiting can be seen with structural gastrointestinal disorders, such as intestinal malrotation with or without intermittent volvulus and intussusception, in which obstruction can be intermittent; however, such conditions are almost always accompanied by abdominal pain, distention, and bilious vomiting (although this can be a later finding in intussusception), which were not present in this case. Proximal obstructive processes that cause nonbilious vomiting, such as upper duodenal webs or stenosis and gastric-outlet obstruction, are rare. The cyclic vomiting syndrome can cause recurrent and often stereotypical vomiting events with return to health between episodes[4] but is rarely diagnosed in infants and toddlers and should not be considered in patients in these age groups until organic causes have been fully ruled out. Again, none of these conditions can explain the patient's macrocephaly and developmental regression.

METABOLIC DISORDERS

As I mentioned previously, the cyclical pattern of vomiting in this patient makes a nongastrointestinal cause more likely than a gastrointestinal cause. Acute hydronephrosis due to intermittent ureteropelvic-junction obstruction can cause severe intermittent vomiting in a young child and bears consideration in this case, but it cannot explain the macrocephaly. Addison's disease and other causes of adrenal insufficiency can result in recurrent vomiting and lethargy but are usually accompanied by hypotension, hyponatremia, and hyperkalemia, which are not present in this case. The normal blood glucose levels before fluid resuscitation in this patient are not consistent with a diagnosis of diabetic ketoacidosis. Toxic ingestion needs to be considered as a potential cause of vomiting in young children who are becoming more mobile and independent and could also potentially explain the developmental regression seen in this patient, but it would rarely recur unless the ingestion is not acciden-

tal. In addition, toxic ingestion alone would not explain the rapidly growing head circumference, and the urine toxicology screening was negative for toxins.

INBORN ERRORS OF METABOLISM

Patients with metabolic disorders and inborn errors of metabolism commonly present with episodic cyclical vomiting and lethargy during times of stress or exposure to the specific substrate that cannot be metabolized. Many of these disorders have milder forms, in which the presentation is delayed until beyond early infancy, that are associated with developmental regression.[5,6] The laboratory findings obtained during this patient's most recent presentation are notable for low total and free carnitine levels and elevated serum lactate and pyruvate levels, findings that are consistent with but not specific for certain metabolic conditions. However, the absence at each presentation of hypoglycemia and significant acidemia (with the only biochemical evidence of acidemia present during the second vomiting episode) and the normal serum ammonia and amino acid levels and normal acylcarnitine profile make some types of inborn errors of metabolism (e.g., fatty-acid oxidation disorders, disorders of carbohydrate metabolism, organic acidemias, and urea-cycle defects) very unlikely.[5,6]

Patients with mitochondrial disorders may present with no acidosis or hypoglycemia and with high serum lactate levels, as did this patient. However, in most mitochondrial disorders, with the exception of pyruvate dehydrogenase deficiency, the ratio of serum lactate to pyruvate far exceeds the ratio seen in this patient (9:1)[7] and macrocephaly would not be expected. Of all the metabolic conditions, Canavan's disease and certain lysosomal storage diseases most often result in macrocephaly. Although these conditions remain diagnostic possibilities and are further supported by the patient's developmental regression, lysosomal storage diseases in particular are unlikely in this patient because of the absence of organomegaly and of classic facial features. Thus, although the abnormal laboratory findings lend some support to a metabolic derangement, no single metabolic condition fits the clinical presentation perfectly.

NEUROLOGIC CAUSES

Neurologic causes of recurrent vomiting warrant strong consideration in this case. The most striking clinical features are the history of developmental regression, the rapid increase in head circumference (with the most rapid increase occurring between 9 and 14 months of age), and the presence of frontal bossing and a full and visibly pulsatile anterior fontanelle on physical examination. These findings, along with the clinical presentation of recurrent vomiting episodes, support a diagnosis of increased intracranial pressure. In children of this patient's age, causes of elevated intracranial pressure include hydrocephalus, central-nervous-system infections or neoplasm, congenital malformations, and intracranial bleeding, which is often due to accidental or nonaccidental trauma. Given the absence of support for other causes, I suspect that the cause of this patient's symptoms is hydrocephalus.

Dr. Virginia M. Pierce (Pathology): Dr. Morley-Fletcher, would you tell us your impression when you examined the patient?

Dr. Morley-Fletcher: I thought a gastrointestinal cause of this patient's illness was unlikely for several reasons. Although viral gastroenteritis was a consideration, the diarrhea had been only intermittent and the patient had had no sick contacts during any of the episodes. Intolerance of cow's milk protein was considered, but the patient had been able to tolerate dairy in the past and her symptoms persisted even after switching to a soy-based formula. I was unable to fit the abnormal laboratory findings into a typical pattern for any metabolic disease. In addition, the patient had had a normal newborn screening, reportedly normal growth and development until recently, and normal laboratory test results on admission.

My clinical diagnosis was recurrent vomiting caused by increased intracranial pressure, possibly due to hydrocephalus. I met with the patient's parents (her mother was seven months pregnant at that time). I remember how important it was for them to feel that their concerns as parents were heard. My recommendation to them was to seek urgent imaging studies of the head for the patient.

CLINICAL DIAGNOSIS

Increased intracranial pressure, possibly due to hydrocephalus.

DR. JESS L. KAPLAN'S DIAGNOSIS

Hydrocephalus.

IMAGING STUDIES

Dr. Sandra P. Rincon: Magnetic resonance imaging (MRI) of the head, performed at the time of presentation to the other hospital, revealed macrocephaly with marked enlargement of the lateral and third ventricles, a finding that is consistent with hydrocephalus (Fig. 2A and 2D). The fourth ventricle was normal in size. A T_2-weighted image showed abnormal hyperintense signal along the margins of the lateral ventricles that was consistent with transependymal flow of the cerebrospinal fluid, which reflects a component of acute-to-subacute hydrocephalus and an interrupted intraventricular septum; this finding is suggestive of long-standing hydrocephalus (Fig. 2E). A high-resolution T_2-weighted image obtained at this hospital at the time of transfer revealed focal stenosis of the superior aspect of the cerebral aqueduct (Fig. 2B). No abnormal enhancement or signal abnormality was seen that would suggest the presence of an obstructing mass or tumor in the region of the cerebral aqueduct, and there was no evidence of ventriculitis (Fig. 2C). No abnormal susceptibility signal was seen that would suggest previous intraventricular hemorrhage (Fig. 2F). The imaging findings are diagnostic of aqueductal stenosis with hydrocephalus.

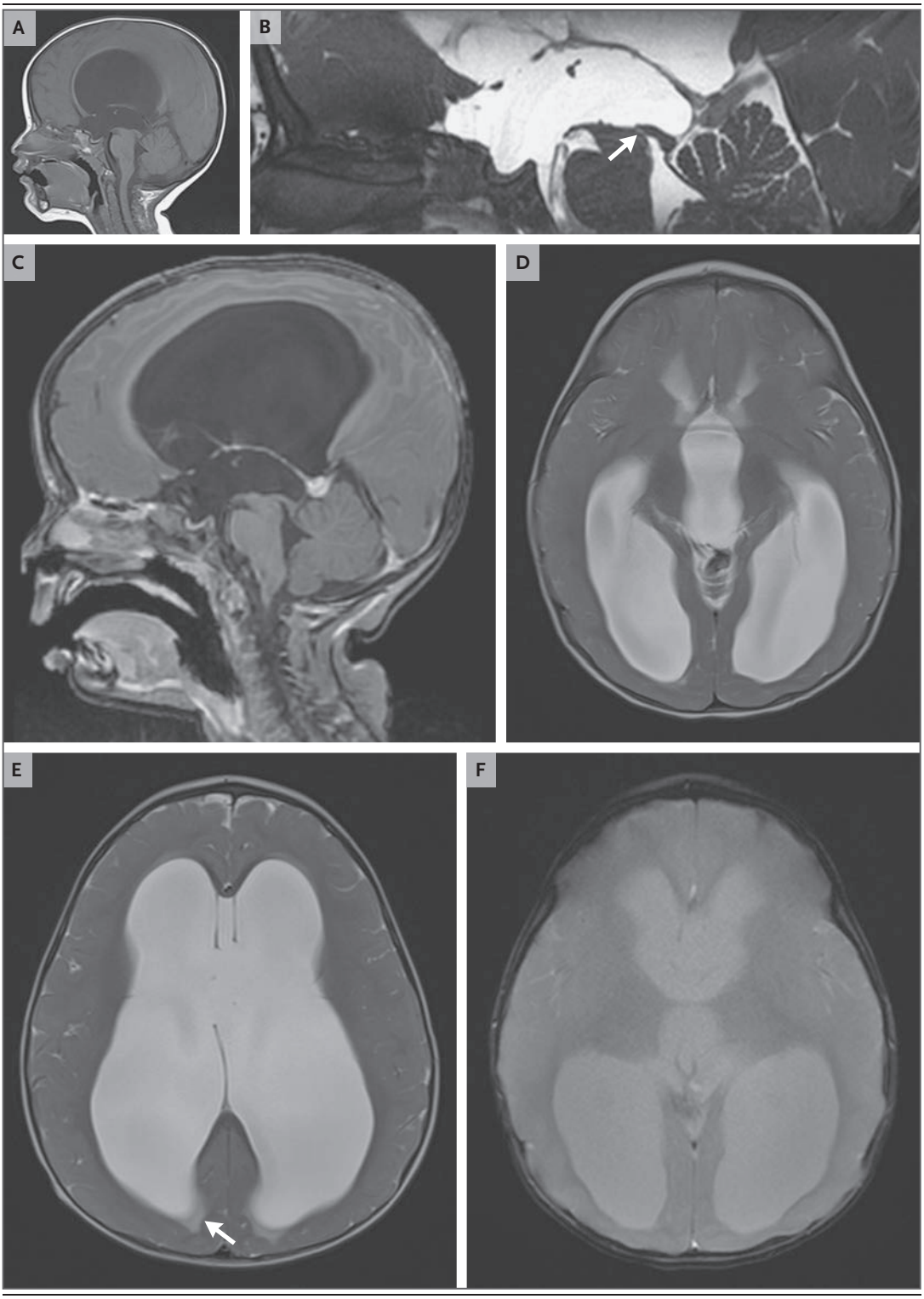
DISCUSSION OF MANAGEMENT

Dr. Ann-Christine Duhaime: This child presented with obstruction of the flow of cerebrospinal fluid at the level of the aqueduct of Sylvius. Since obstruction at this site can be congenital, can be acquired through scarring or inflammation, or can reflect a neoplasm or other expanding lesion, additional high-resolution imaging studies were performed before surgical intervention to assess whether an obstructive lesion that might require specific therapy was present. When no lesion was identified, the target of management became relief of the mechanical obstruction.

The traditional approach to management of aqueductal stenosis is placement of a ventriculoperitoneal shunt, which bypasses the obstruction by draining the extra cerebrospinal fluid from the ventricle into the peritoneal cavity, where it can be absorbed back into the bloodstream. The surgery for initial shunt placement has an excellent safety profile and is associated with a high rate of efficacy in relieving increased intracranial pressure and diminishing ventricular volume. However, shunts carry risks; the two most common are shunt malfunction and shunt obstruction. Shunt malfunction occurs in the majority of children with shunts and can result in acutely increased intracranial pressure for which urgent surgery for shunt repair is required. Children with a shunt placed early in life undergo an average of three or four operations during childhood. Shunt placement is associated with a mortality of approximately 1 to 4%.[8] The other common complication of shunt placement is infection, which occurs in approximately 5 to 10% of shunt operations and has been associated with increased morbidity, including an increased rate of cognitive deficits. Treatment of shunt infection typically requires removal of the shunt and administration of a course of intravenous, and sometimes intrathecal, antibiotic agents.

Advances in endoscopic techniques have made opening the obstructed cerebrospinal fluid pathways feasible. Improvements in optics, miniaturization, and tools for use through the endoscope have led to increased use of endoscopic third ventriculostomy. MRI-guided navigation assists in the selection of the best possible trajectory for endoscope placement and minimizes complications. Endoscopic third ventriculostomy involves inserting the endoscope through a small opening near the coronal suture and advancing it through the frontal cortex, lateral ventricle, and foramen of Monro and into the third ventricle. Fenestration is then performed through the floor of the third ventricle and into the subarachnoid space, thus allowing the cerebrospinal fluid to enter this space and bypassing the obstruction at the aqueduct.

As compared with shunt placement, endoscopic third ventriculostomy as a treatment for hydrocephalus has both advantages and disadvantages.[9] Advantages include the possibility of relieving intracranial pressure without depending on a shunt, which is associated with risks of malfunction and infection. However, some young children who undergo endoscopic third ventriculostomy do not absorb the cerebrospinal fluid sufficiently, so they may continue to have signs and symptoms of active hydrocephalus and may require shunt placement.[10] Although surgical complications are uncommon, they can be serious; possible complications include injury to the basilar artery during fenestration of the third ventricular floor. The fenestration site can close and thus require repeat fenestration or shunt



placement. Reocclusion of the fenestration site is associated with a risk of death, as is shunt malfunction.[11] The rates of long-term success associated with shunts and endoscopic third ventriculostomy are similar; endoscopic third ventriculostomy is associated with a higher rate of initial compli-

Figure 2 (facing page). MRI of the Head Obtained on Presentation.

A sagittal T_1-weighted image (Panel A), obtained without the administration of gadolinium, shows macrocephaly with marked enlargement of the lateral and third ventricles and downward bowing of the floor of the third ventricle into the sella turcica; the fourth ventricle is normal in size. A high-resolution sagittal T_2-weighted image (Panel B) shows stenosis of the cerebral aqueduct (arrow). A sagittal T_1-weighted image (Panel C), obtained after the administration of gadolinium, shows no abnormal enhancement; there is no evidence of an obstructing mass or ventriculitis. Axial T_2-weighted images (Panels D and E) show transependymal flow of the cerebrospinal fluid along the margins of the lateral ventricles (Panel E, arrow) and an interrupted intraventricular septum, a finding that can be seen with long-standing hydrocephalus. A susceptibility-weighted image (Panel F) shows no evidence of intracranial hemorrhage.

cations but also with a higher rate of long-term success.[9]

For all these reasons, the decision of whether to proceed with shunting or endoscopic third ventriculostomy in this patient required careful counseling and consideration of the options with the family. In elective cases, this can be done over several meetings, but in this case, the child had progressive symptoms and clear neurologic decline and needed prompt intervention. Appropriate counseling requires care and sensitivity to the fact that families may feel overwhelmed by decisions involving technical and often frightening information. In this case, the patient's parents were open and able to participate in a discussion of the alternatives and felt comfortable opting for endoscopic third ventriculostomy as an initial approach, with shunt placement reserved for a second option if the anatomy appeared to be unfavorable for endoscopic third ventriculostomy or if endoscopic third ventriculostomy was not effective.

In this young child with a large head circumference, we took several approaches to minimizing the complications of endoscopic third ventriculostomy. First, the entry point for the endoscope was carefully planned to create a straight trajectory through the foramen of Monro and toward an appropriate fenestration target point in the floor of the third ventricle to minimize manipulation or damage to the surrounding fornices, which are important for memory function. Second, the child's head was positioned with the entry point most superior and parallel to the floor to minimize the loss of cerebrospinal fluid from the enlarged lateral ventricle, which can lead to rapid shifts in fluid and subdural bleeding. Third, a curvilinear incision was made to provide access to the burr hole without directly overlying it to minimize the risk of a postoperative cerebrospinal fluid leak. Fourth, after opening of the dura, the pia mater was coagulated and incised, and entry into the ventricle was performed gently and slowly along a predetermined path, to minimize the risk of parenchymal hemorrhage (Video 1, available with the full text of this article at NEJM.org). Fifth, fenestration was performed in a controlled fashion, with gentle, focused handling of tissue with blunt instruments. Great care was taken to avoid damage to the basilar artery and its branches, as well as injury to the walls of the third ventricle, which can affect endocrine and other functions. The fenestration was enlarged gradually by means of inflation of a balloon catheter. Finally, after ascertaining that fenestration was complete by advancing the endoscope into the subarachnoid space and ensuring that all membranes had been opened, the endoscope was carefully withdrawn, the cortex was inspected, and careful closure was performed to minimize the chance of postoperative cerebrospinal fluid leak and infection. The procedure was completed without complications.

A video showing a portion of the endoscopic third ventriculostomy is available at NEJM.org

Dr. Rincon: MRI of the head, performed after the endoscopic third ventriculostomy, revealed that the size of the ventricles had decreased (Fig. 3A and 3C). The previously seen transependymal edema had resolved, and the sulci were better defined because of the decreased mass effect. There was persistent narrowing of the cerebral aqueduct.

FOLLOW-UP

Dr. Morley-Fletcher: After the procedure, the patient was more alert. No new focal deficits or seizure-like activity developed. She was discharged home on the second postoperative day.

The patient was seen by early intervention services, the physical therapy service, and the occupational therapy service. She had rapid improvement in her level of alertness and interest in reaching for objects and gradual improvement in motor skills. Subsequent imaging studies showed improvement in hydrocephalus, and the

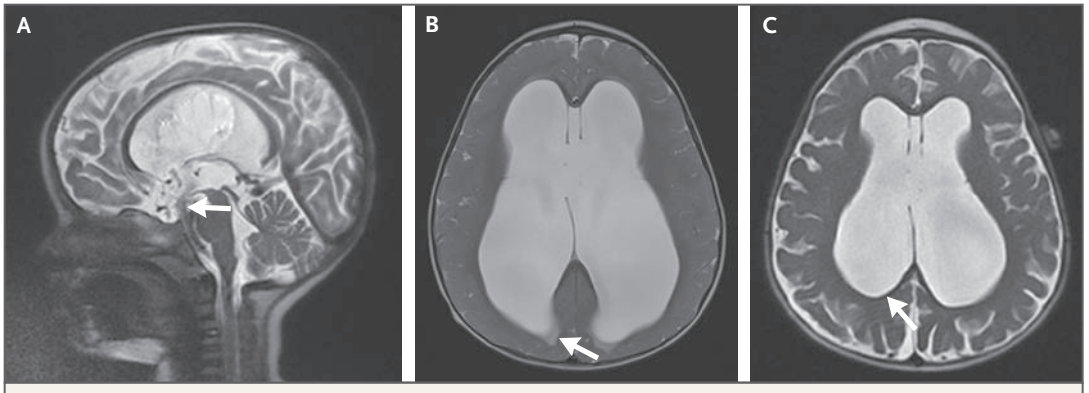


Figure 3. MRI of the Head Obtained after the Endoscopic Third Ventriculostomy.

A sagittal T_2-weighted image (Panel A) shows postsurgical changes related to endoscopic third ventriculostomy for treatment of hydrocephalus. There is flow-related signal in the region of the anteroinferior third ventricle that extends into the suprasellar cistern, a finding that is consistent with patency of the ventriculostomy (arrow). Axial T_2-weighted images obtained before and after the ventriculostomy (Panels B and C, respectively) show a decrease in the size of the lateral ventricles and resolution of transependymal edema after surgery (arrows); the sulci are better defined on the postoperative image because of the decreased mass effect.

growth of her head circumference slowed. One year after surgery, the patient still has some unsteadiness of gait but is meeting most neurodevelopmental milestones.

Seeing the dramatic improvement and resolution of the patient's symptoms over time was a rewarding experience for me as a physician in training. I have come to get to know the patient and her parents very well during their journey.

Dr. Pierce: We are privileged to have the patient and her parents here. I would like to ask her mother to say a few words about their experience of their daughter's illness.

The Patient's Mother: Our daughter was admitted to the hospital on the evening of a major holiday. Dr. Morley-Fletcher asked us, "What is she like when she's not sick?" I said that she was very outgoing and seemed to have a magnetic personality — wherever we went, people were drawn to her. I was afraid that the MRI would show a brain tumor. Once we knew that she needed to have brain surgery, I was nervous about that. We are so thankful that everything went well. It has been a challenging year for us, since we have had to take her to so many appointments. But the other day, as I was making a follow-up appointment on our way out of physical therapy, I watched her interact with an elderly woman in the waiting room, and I could see that talking with my 2-year-old daughter was making this woman so happy. She brings joy to everyone around her. We wouldn't want anything about her to be different.

FINAL DIAGNOSIS

Aqueductal stenosis with hydrocephalus.

This case was presented at the postgraduate course, "Primary Care Pediatrics," codirected by Peter Greenspan, M.D., John Patrick T. Co, M.D., M.P.H., Ronni L. Goldsmith, M.D., Janice A. Lowe, M.D., and Benjamin A. Nelson, M.D., and sponsored by the Harvard Medical School Department of Continuing Education.

Disclosure forms provided by the authors are available with the full text of this article at NEJM.org.

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