

# Appendicitis in a Child with Paraplegia

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**T**he assessment and diagnosis of acute abdominal disorders are particularly challenging with regard to patients with paraplegia secondary to spinal cord lesions.<sup>1,2</sup> Because of the high morbidity and mortality rates associated with acute abdominal disorders in this population (30% to 50% and 10% to 15%, respectively),<sup>1,3</sup> timely and accurate diagnosis is crucial. Impairment of motor and sensory function below the level of the spinal cord lesion complicates the evaluation of these patients.<sup>1</sup> This article describes the presentation and clinical course of a child with paraplegia who developed appendicitis. The pathophysiology and general clinical features of an acute abdomen in a patient with paraplegia are discussed.

### CASE PRESENTATION

#### Initial Presentation

A 9-year-old boy with paraplegia and a history of spina bifida, scoliosis, and a ventriculoperitoneal (VP) shunt presented to our pediatric clinic with the chief complaint of fever with chills and sweats for 3 days; during that time, his body temperature had been as high as 103°F. He had been examined in the pediatric emergency department (ED) on the first day that he experienced a fever. At that time, he also had a headache, felt fatigued, and had loss of appetite; these symptoms began the day before his visit to the ED. The results of a shunt series and a computed tomographic (CT) scan of his head, obtained during his ED visit, were within normal limits. An abdominal radiograph showed evidence of increased stool. He was administered an enema and was discharged with a diagnosis of constipation and fever. He was instructed to follow up with his pediatrician if his fever persisted beyond 48 hours, which led to his presentation at our clinic.

By the time he came to our clinic, his loss of appetite, headache, and feelings of fatigue had lasted for 4 days. The day before presentation, he had 2 loose stools, which were green, nonbloody, and nonmucoid. He had no runny nose, cough, or shortness of breath. He stated that he had left ear pain that started on the day of presentation. He complained of an uneasy feeling, which he described as "butterflies fluttering" in his

left upper abdomen. He had experienced no vomiting episodes or changes in urine output. He had not been in contact with ill individuals and had no history of travel. There was no significant family history.

#### History

The patient was born full term by spontaneous vaginal delivery. He had a high meningomyelocele, spina bifida, hydrocephalus, and scoliosis. The VP shunt was placed when he was 2 days old. The VP shunt was subsequently revised 5 times, the last revision occurring 2 years ago. He had a bilateral inguinal hernia repair and orchidopexy 8 years ago. He was allergic to latex, was wheelchair bound, lived at home with his parents, and attended regular school, where he was an exceptional student.

#### Physical Examination

The patient was alert and active and in no apparent distress. His body temperature was 104.5°F; pulse, 150 bpm; respiratory rate, 30 breaths/min; and blood pressure, 127/34 mm Hg. He was very friendly and talkative, complaining that he "hated coming to the hospital." His mucous membranes were moist, and his pharynx was very mildly injected, with tonsils of normal size. The right tympanic membrane was normal. However, the left tympanic membrane was minimally erythematous but had normal mobility with easily identifiable landmarks. His VP shunt reservoir appeared patent by palpation. His neck was supple. His lungs were clear, and his heart sounds were normal, with no murmurs. His abdomen was soft, obese, and nondistended, with no mass or organomegaly; there was no tenderness, rigidity, or guarding. On deep palpation of the right lower quadrant of the abdomen, the patient demonstrated a flexion reflex at the right hip joint. His hands and feet were cold and clammy, and he had beads of sweat on his forehead. Pulses were brisk. His genitalia and extremities were normal. A neurologic

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examination revealed bilateral flaccid lower extremities with no sensation below the level of the umbilicus. His shunt was patent, and the shunt tract was not erythematous or swollen.

#### Laboratory Studies

A complete blood count yielded a leukocyte count of  $18.8 \times 10^3/\mu\text{L}$  (85% neutrophils, 9% lymphocytes), a hemoglobin concentration of 11.6 g/dL, a hematocrit of 34.5%, a platelet count of  $349.9 \times 10^3/\mu\text{L}$ , a erythrocyte sedimentation rate of 103 mm/h, and a C-reactive protein concentration of 31 mg/dL. The results of a urine analysis were normal. Blood and urine cultures sent to the laboratory for evaluation did not show any bacterial growth.

#### Radiographic Studies

A shunt series showed a 2-cm linear calcified density of uncertain significance in the right lower abdominal area. An abdominal sonogram was obtained to help rule out an infectious cyst. It showed some free fluid in the peritoneal cavity. The patient was admitted to the pediatric floor of the hospital for observation and for further investigation of the source of fever. A chest radiograph showed no abnormalities. A CT scan of the abdomen was obtained and showed evidence consistent with a ruptured appendix, with abscess formation. The VP shunt traveled directly through the phlegmon.

#### Management

The patient was placed on intravenous hydration therapy and was intravenously administered clindamycin and cefotaxime. An appendectomy was performed, with externalization of the VP shunt. The VP shunt was externalized to reduce the risk of shunt infection and to help in postoperative recovery. During the procedure, the surgeons discovered a retrocecal ruptured gangrenous appendix with a freely floating fecalith.

The patient remained febrile for the first 5 days after the surgery. During the postoperative period the surgical wound culture grew pan-sensitive *Escherichia coli* bacteria and also *Pseudomonas aeruginosa* bacteria that were sensitive to gentamicin and ceftazidime. The cefotaxime was subsequently discontinued, and the patient was started on ceftazidime. Forty-eight hours after the change of antibiotics, he was afebrile. (He had been treated with intravenous antibiotics for a total of 10 days.) The VP shunt was reinternalized on the 12th day of admission, after he was asymptomatic for 2 days without antibiotic therapy. His oral feeding was advanced over 2 days.

#### Outcome

The patient was discharged on the 14th day after hospital admission. He was evaluated in the pediatric clinic on the day after discharge and was afebrile. He appeared well and was still very talkative. His C-reactive protein level had decreased from 31 to 1 mg/dL. His leukocyte count had also diminished.

#### DISCUSSION

##### Nerve Disruptions Associated with Paraplegia

Visceral and somatic sensory nerves that transmit pain and give rise to other symptoms and signs of an acute abdomen are disturbed in patients with paraplegia.<sup>4</sup> However, the disturbance is not necessarily complete. Some patients with paraplegia continue to experience some degree of somatic sensation (eg, pain) below the level of their spinal cord lesion (although intermittently), and in many patients, visceral sensation is largely preserved. Several explanations for the preservation of visceral sensation in these patients have been suggested.<sup>1,5</sup> One explanation is that some visceral sensory fibers may not be affected by a lesion because they may climb along the autonomic ganglia before they enter the spinal cord.<sup>6</sup> Also, some visceral sensory fibers may travel with the vagus nerve, circumventing the cord lesion, and some fibers may travel undamaged through the injured area.<sup>1</sup> (The actual mechanism may be a combination of these hypotheses.) Aggravated spasticity of the muscles of the abdominal wall and lower limbs may be attributable to irritation of anterior horn cells. This irritation occurs because of augmented visceral sensations secondary to visceral distention.<sup>7</sup> Provided the reflex arc is intact, there will be reflex activity below the level of the lesion.<sup>1,8</sup>

##### Patient Presentation

The systemic symptoms and signs of appendicitis may be subtle early in the disease process in patients with paraplegia. Among patients with spinal cord lesions, certain clinical features can suggest acute abdominal disease. Some of these features are anorexia, nausea, fatigue, abdominal distention, vague abdominal pain, and increased spasticity of abdominal or lower-limb muscles.<sup>1-4,6,7,9-14</sup> A patient may also experience autonomic dysreflexia, which may cause diaphoresis, tachycardia or bradycardia, hypertension, arrhythmias, visual changes, and headaches. He or she may indicate a feeling of restlessness and that "something does not feel right" in his or her abdominal area but most likely would not be able to pinpoint the sensation.<sup>1</sup>

There are relatively few afferent fibers associated with the viscera, and the sense of pain is not associated

with many visceral organs. However, as Wenger indicated,<sup>15</sup> even vague internal sensations are enough for humans to understand that they may have an internal disorder. Such vague sensations are often described as “lumps in the throat,” “a sinking feeling,” “butterflies in the stomach,” and “a feeling of heaviness or lightness.”<sup>15</sup>

A flexion reflex at the hip joint on deep palpation of the abdomen, without pain, has been observed in patients with paraplegia who have abdominal diseases. Such a reflex was observed with regard to our patient. The physiologic mechanisms that come into play regarding the withdrawal of the legs by flexion are unclear; pathology of the legs (eg, pressure ulcers or ingrown toenails) as well as intra-abdominal processes can change the balance between the stretch and flexion reflexes.<sup>8,16</sup> Flexion reflex relates to the motion corresponding to withdrawal from a noxious stimulus.<sup>16</sup> In patients with paraplegia, the flexion reflex can serve as a warning that something has interfered with the balance between flexion and stretch reflexes.<sup>8,16,17</sup> Spasticity of the muscles of the abdomen or lower limbs may worsen in some patients.<sup>7</sup>

Patients with spinal cord lesions at or above thoracic vertebra 6 (T6) with acute abdominal pathology may present with autonomic dysreflexia (AD). This can occur because of the distention or perforation of a hollow organ<sup>2,9</sup> and can be extremely dangerous. Lesions at or above T6 interrupt control over major sympathetic nerve tracts in the thoracolumbar region.<sup>3</sup> In patients with AD, deleterious visceral or cutaneous stimuli can cause an uncontrolled and sudden reflex sympathetic reaction.<sup>4</sup> An associated fever is usually attributable to the abdominal disease process rather than to AD. Early recognition of this potentially life-threatening condition can aid in the diagnosis of acute abdominal surgical emergencies.

Coexisting morbidity may complicate the clinical picture of an acute abdomen in this patient population. Chronic urinary tract infections, gastroparesis, decubitus ulcers, pharyngitis, otitis media, pneumonia, and upper respiratory infections are important diagnoses that may present simultaneously and interfere with efforts to establish an accurate diagnosis.<sup>4</sup>

### Differential Diagnosis

Pyelonephritis is usually associated with high fever, bacteria and leukocytes in the urine, and positive results on urine culture testing for bacteria. Patients with this disease usually demonstrate a prompt and impressive response to antibiotic therapy, provided there is no accompanying gram-negative sepsis or septic shock.<sup>7</sup> It is important to distinguish pyelonephritis

from other causes of an acute abdomen, because patients with paraplegia can have pyuria without genitourinary infections. This finding could falsely divert one's attention to the urinary system.

Hematuria and crystalluria may indicate renal stones; an abdominal radiograph or renal sonogram can aid in the diagnosis.<sup>7</sup> Fecal impaction in patients with paraplegia can present either as constipation or diarrhea. It may occur with fever, nausea, vomiting, abdominal pain, abdominal distention, and autonomic hyperreflexia. Findings on the physical examination may include an abdominal fecal mass and impacted feces in the rectum. Immediate management would include bowel decompression.<sup>7</sup>

As highlighted by the clinical course of the patient in this report, an important and life-threatening diagnosis could be acute appendicitis. A helical CT scan and diagnostic laparoscopy are very helpful investigations. Also, it is important to rule out intra-abdominal abscesses. Rupture of the urinary bladder should be considered if tenderness is suprapubic. Splenic rupture can be rapidly fatal and should be kept in mind in a patient with a history of trauma, rapidly worsening anemia, or shock.<sup>7</sup> If the clinical findings point to an upper abdominal process, it is important to exclude cholecystitis, cholelithiasis, peptic ulcers, pancreatitis, and hepatitis.<sup>7</sup>

A fever could result from soft tissue infections attributable to pressure ulcers or osteomyelitis. Also, patients with paraplegia are prone to respiratory complications, such as pneumonia and pulmonary embolism, which could cause clinical features mimicking an acute abdomen.

A cerebrospinal fluid pseudocyst can produce signs and symptoms of an acute abdomen in children with a VP shunt. It can be diagnosed through a shunt series or a CT scan of the abdomen and usually presents in the upper abdominal quadrants. A shunt infection may also result in an acute abdomen. This is managed by antibiotic therapy and removal of the shunt from the abdominal cavity with its conversion into a ventriculoatrial shunt.<sup>18</sup>

### Diagnostic Methods

The majority of patients with paraplegia occasionally have pyuria, usually after undergoing catheterization procedures. Approximately 45% of patients with spinal cord lesions who have intra-abdominal pathology may not have an elevation or left shift in their leukocyte count.<sup>1</sup> Therefore, their nonspecific clinical features may be assumed to arise from the genitourinary system. For this reason, laboratory tests are not very reliable.<sup>1,2</sup> Neumayer

indicated that radiographic studies were the most helpful methods of reaching a correct diagnosis.<sup>1</sup> Abdominal radiographs, chest films, oral cholecystograms, ultrasonographic evaluations, CT scans, intravenous pyelograms, and barium studies led to the correct diagnosis in 77% of the patients followed in this study.<sup>1</sup> The results of an ultrasonographic evaluation are, however, operator dependent; one has to rely heavily on the skill of the technician performing the examination.<sup>19</sup>

Focused helical CT scans can be faster and more accurate than the previously mentioned methods.<sup>20</sup> Findings suggestive of appendicitis are the presence of an abscess or phlegmon, nonfilling of the appendix with contrast material, an enlarged or thickened appendix, periappendiceal air, an appendicolith, and periappendiceal fat stranding.<sup>19,20,21</sup> Diagnostic laparoscopy provides a definitive diagnosis if the clinician has a high index of suspicion despite normal results on laboratory and radiologic investigations. It has the added advantage of being therapeutic if appendicitis is diagnosed and an appendectomy is to be performed.

#### General Patient Outcomes

According to Strauther et al,<sup>4</sup> there is a significant time lag from when symptoms first develop to the time the patient is correctly diagnosed. In this study, the average length of time that symptoms persisted before patients were admitted to the hospital was 4 days. This could be attributable to a lack of understanding by the patient or caregiver of the implications of the clinical picture. Only 35% of the patients in this study were diagnosed with appendicitis on hospital admission.<sup>4</sup> The average interval of time the patients were hospitalized prior to diagnosis was 2 days, demonstrating the challenges such a case presents to the clinician. Perforation of the appendix was found during surgery in 92% of patients evaluated.<sup>4</sup> The average length of inpatient stay was 16 days.<sup>4</sup> The mortality rate attributable to an acute abdominal process in patients with paraplegia ranges from 10% to 15%.<sup>3,9</sup>

#### CONCLUSION

Knowledge of the pathophysiology of acute abdominal conditions in children with paraplegia can help in the early recognition, diagnosis, and management of the abdominal disorders. Abnormal pain perception and concomitant chronic diseases can make the diagnosis of abdominal complaints challenging with regard to a patient with paraplegia.<sup>1</sup> The physician caring for such a patient should be familiar with the altered physiologic response the patient may have to the pathologic condition. Early recognition of the unusual clinical

signs and symptoms of an acute abdomen could lead to a timely diagnosis.

A high index of suspicion must be maintained when investigating fever of uncertain origin in patients with paraplegia, especially those in the pediatric age group. Children may not be able to adequately describe their symptoms. They may also have coexisting conditions that may cause fever, including pharyngitis, otitis media, urinary tract infection, or upper respiratory infection. Patients with paraplegia who have complaints of abdominal pain or tenderness or gastrointestinal symptoms must be thoroughly investigated to rule out life-threatening pathology. A focused helical CT scan is highly sensitive in diagnosing appendicitis and can help one to make a diagnosis promptly. **HP**

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