CASE REPORT

Interval Laparoscopic Appendectomy for Appendicitis Complicated by Pylephlebitis

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ABSTRACT

Background: Although rare, portal mesenteric venous thrombosis and pylephlebitis remain potential life-threatening sequelae of ruptured appendicitis in children. Treatment recommendations from recent reports have included urgent exploratory laparotomy with appendectomy, prolonged intravenous antibiotic therapy, and anticoagulation for up to a year.

Methods: This report describes successful management of pylephlebitis and mesenteric venous thrombosis complicating ruptured appendicitis with intravenous antibiotics and anticoagulation followed by interval laparoscopic appendectomy.

Results: A previously healthy 5-year-old girl was diagnosed with ruptured appendicitis complicated by pylephlebitis and mesenteric venous thrombosis at the time of presentation. She was treated with intravenous antibiotics and anticoagulated for 3 months. She subsequently underwent interval laparoscopic appendectomy. At 3-year follow-up, she is healthy without evidence of adverse sequelae.

Discussion: This is the first reported case of successful, minimally invasive management of ruptured appendicitis complicated by mesenteric venous thrombosis and pylephlebitis.

Conclusion: Similar treatment of other children with this rare presentation seems reasonable.

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INTRODUCTION

Mesenteric venous thrombosis and pylephlebitis are rare sequelae of ruptured appendicitis in children. Traditional management has included urgent exploratory laparotomy with appendectomy, intravenous antibiotics, and long-term anticoagulation. A recent review of the English-language literature revealed no published reports of management of appendicitis complicated by pylephlebitis with minimally invasive surgical techniques. We describe successful management with intravenous antibiotics and anticoagulation followed by interval laparoscopic appendectomy in a 5-year-old girl.

CASE REPORT

A previously healthy 5-year-old girl presented with a 7-day history of fever, vomiting, and anorexia and a 24-hour history of diffuse abdominal pain. At the time of presentation, her rectal temperature was 104.1°F. Physical examination demonstrated generalized voluntary abdominal guarding but no focal tenderness. Notable laboratory abnormalities included white blood count 14.5 ($x10^9$/L) with 83% polymorphonuclear leukocytes, aspartate amino transferase (AST) 66 U/L, amino alanine transferase (ALT) 107 U/L, alkaline phosphatase 267 U/L, and total bilirubin 1.1 mg/dL. Computed tomography (CT) of the abdomen and pelvis was obtained and revealed a thickened retrocecal appendix with associated inflammatory changes but no focal fluid collection **(Figure 1),** thrombosis of the superior mesenteric vein **(Figure 2)** with extension of the thrombus into the left branch of the portal vein, and impaired perfusion of the left lobe of the liver **(Figure 3).**

The patient was admitted to the hospital, hydrated, placed on broad-spectrum intravenous (IV) antibiotics, and anticoagulated with IV unfractionated heparin. Her fevers, nausea, and pain quickly resolved. A diet was offered on hospital day 4, which she tolerated without difficulty. On hospital day 6, a follow-up CT scan was obtained and dem-

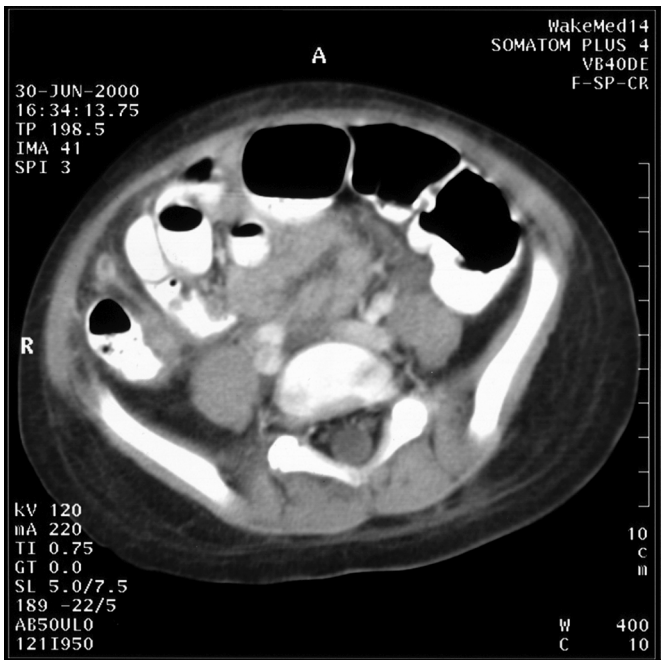

Figure 1. Thickened appendix with associated inflammatory changes.

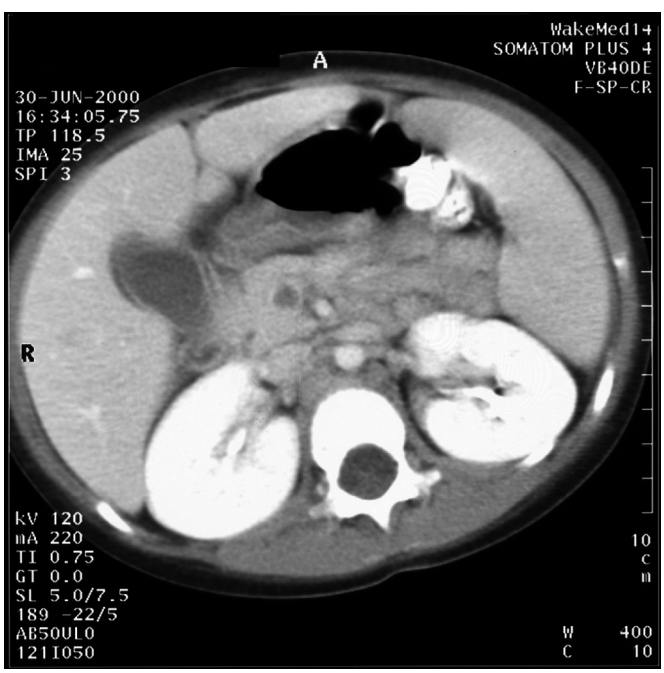

Figure 2. Thrombosis of the superior mesenteric vein.

onstrated dramatic improvement in hepatic perfusion **(Figure 4),** despite residual superior mesenteric vein thrombus. After 12 days of IV antibiotics and heparin, she was discharged home. Discharge medications included low-molecular weight heparin, enoxaparin 40 mg sq qd (Lovenox, Aventis Pharmaceuticals, Inc, France). She was subsequently converted to oral warfarin, dose-adjusted to maintain an international normalized ratio (INR) between 2.0 and 3.0.

Pediatric hematology consultation was obtained, and a hypercoagulability workup was initially consistent with heterozygous protein C deficiency: activity 35% (normal, 64% to 126%). Antithrombin III level was also mildly low at 60% (normal, 70% to 115%). Protein C antigen level while the patient was on heparin was 99% (normal, 71% to 157%). However, coagulation functional screens obtained at the time of clot presentation are known to be unreliable. After 3 months, the warfarin was discontinued; repeat testing at that time revealed no evidence of protein C deficiency (activity 82% of normal) nor of antithrombin III deficiency (101% of normal).

Four months after initial presentation, the patient underwent laparoscopic appendectomy. Intraperitoneal examination revealed mild scarring and evidence of previous inflammation, but no other abnormality **(Figure 5).** The appendix was removed without difficulty by using a 3-trocar technique. Operative time was 68 minutes. Subsequent histologic examination of the appendix revealed fibrosis and evidence of prior sealed perforation. She had an uncomplicated postoperative course and was discharged home on the third postoperative day. She is now 3 years postappendectomy with no evidence of long-term sequelae. Notably, she has had no further thrombotic events.

DISCUSSION

Before the second half of the 20th century, the incidence of mesenteric venous thrombosis and pylephlebitis complicating appendicitis was less than 1%.[1] Since the advent of modern antibiotic therapy and aggressive surgical approaches to appendicitis, pylephlebitis has become an exceedingly rare, yet still potentially fatal, complication of appendicitis. Although the low incidence makes it difficult to know the true mortality rate of pylephlebitis secondary to appendicitis, modern estimates of the mortality of pylephlebitis from all causes are as high as 50%.[2,3] Although pylephlebitis may rarely occur shortly after the onset of symptoms, it most often is associated with cases of appendicitis in which a delay occurs in presentation or diagnosis. Alternatively, pylephlebitis occasionally occurs after appendectomy in cases of ruptured appendicitis. Because the presentation of appendicitis can be atypical in young children, they are at increased risk for delayed presentation to a health care provider and diagnosis. Consequently, children may be particularly vulnerable to pylephlebitis.

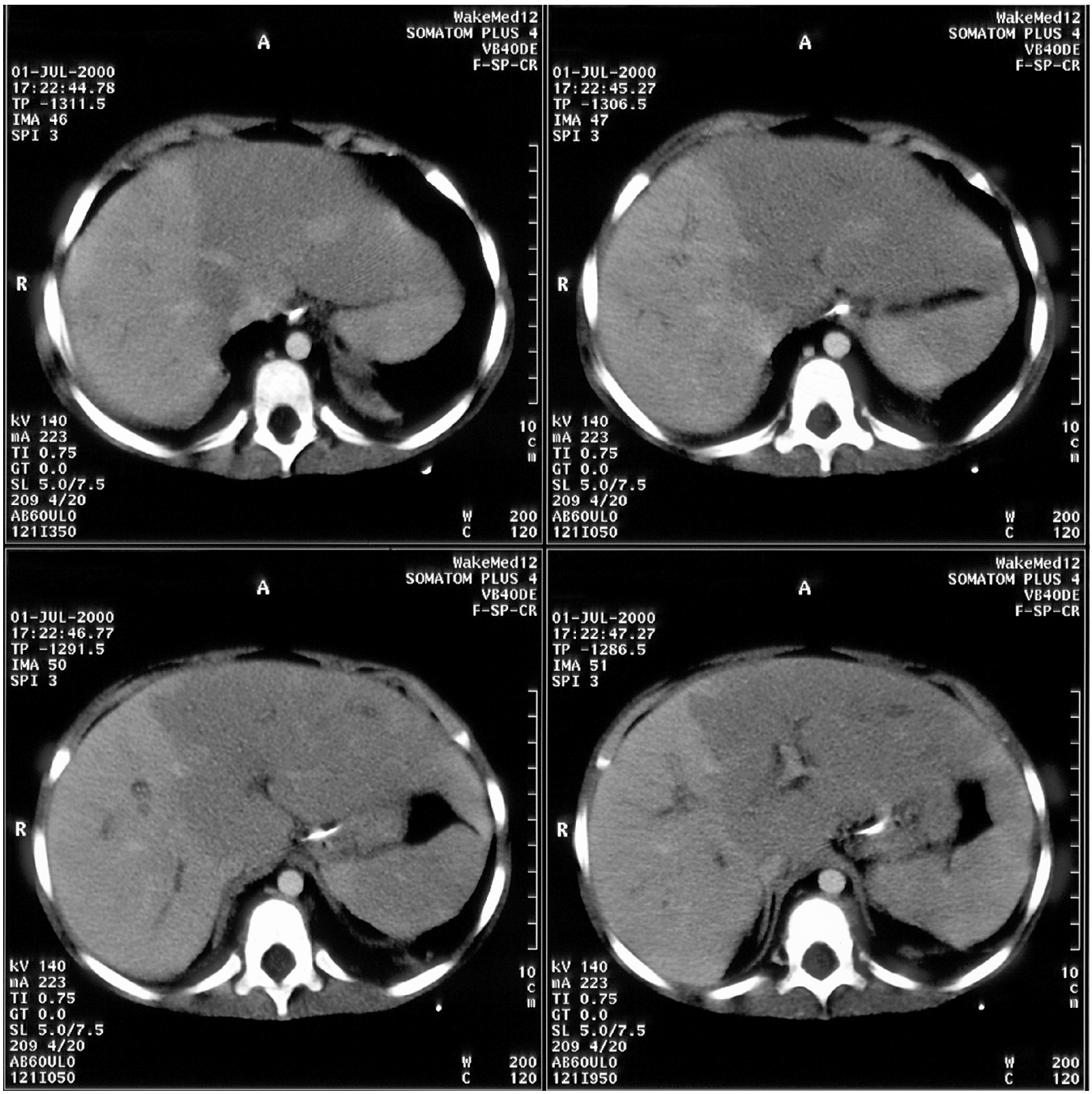

Figure 3. Impaired perfusion of the left lobe of the liver, secondary to extension of thrombus into the left branch of the main portal vein.

Traditional management of appendicitis complicated by pylephlebitis has included intravenous antibiotic therapy and emergent exploratory laparotomy with appendectomy. The need for long-term anticoagulation has been a topic of debate for the last century and remains disputed.[2–6] A review of the English-language literature by using PubMed (National Center for Biotechnology Information, Bethesda, MD) yielded 24 articles published since 1959, describing 34 cases of mesenteric venous thrombosis and pylephlebitis complicating appendicitis. Additionally, a

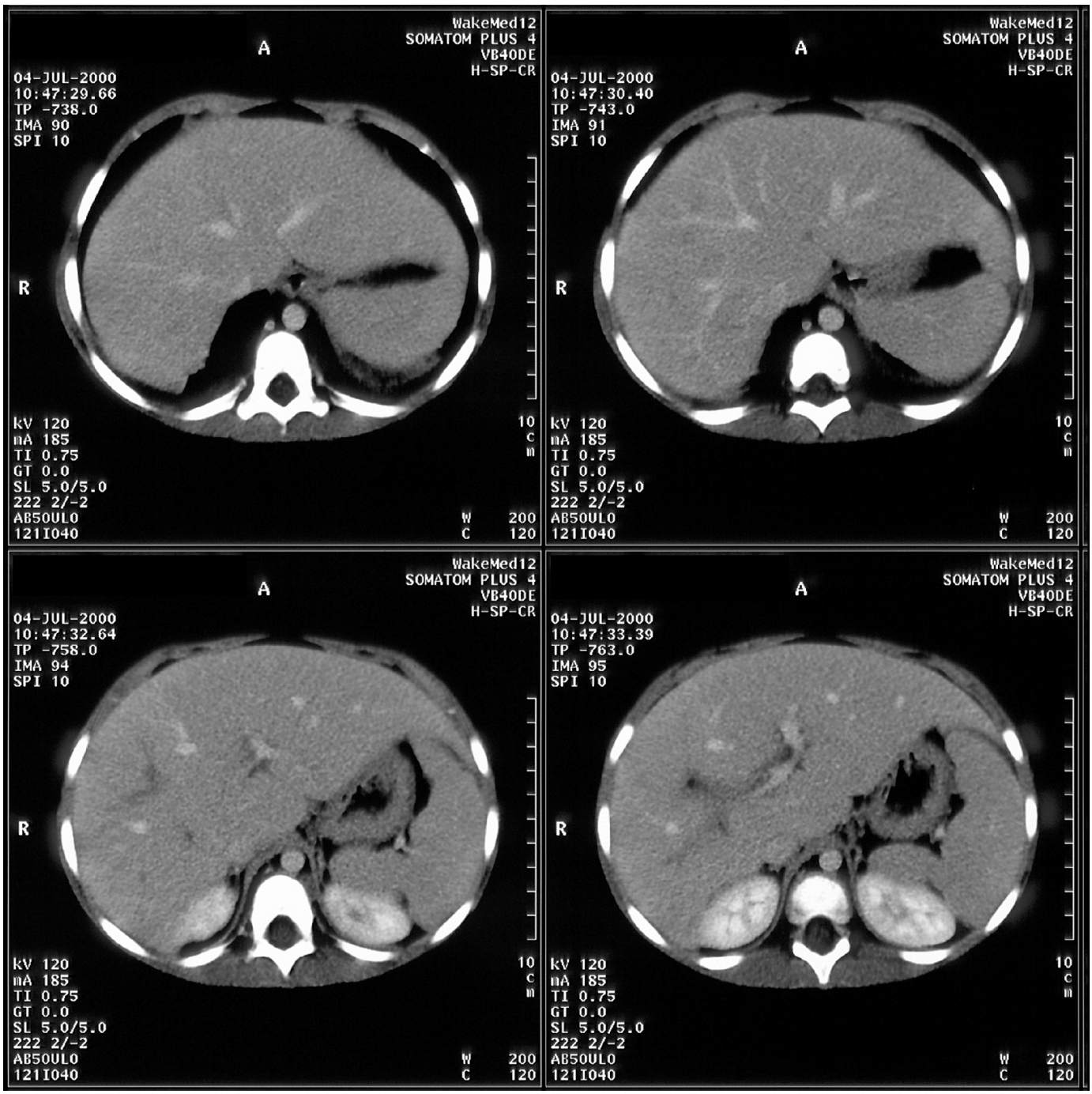

Figure 4. Improved perfusion of the liver after 6 days of intravenous antibiotics and anticoagulation.

review by Klinefelter et al[7] details 62 cases reported between 1926 and 1959. Of the 34 reported cases since 1959, only 4 occurred in females. This is consistent with Klinefelter's observation that roughly 85% of cases of pylephlebitis secondary to appendicitis occur in males.[7] Forty-four percent (15/34) of the cases occurred in children **(Table 1).** In the children, pylephlebitis was present at the time of diagnosis of appendicitis in 10 cases. Seven of these patients were treated with urgent exploratory laparotomy and appendectomy, and several of these pa-

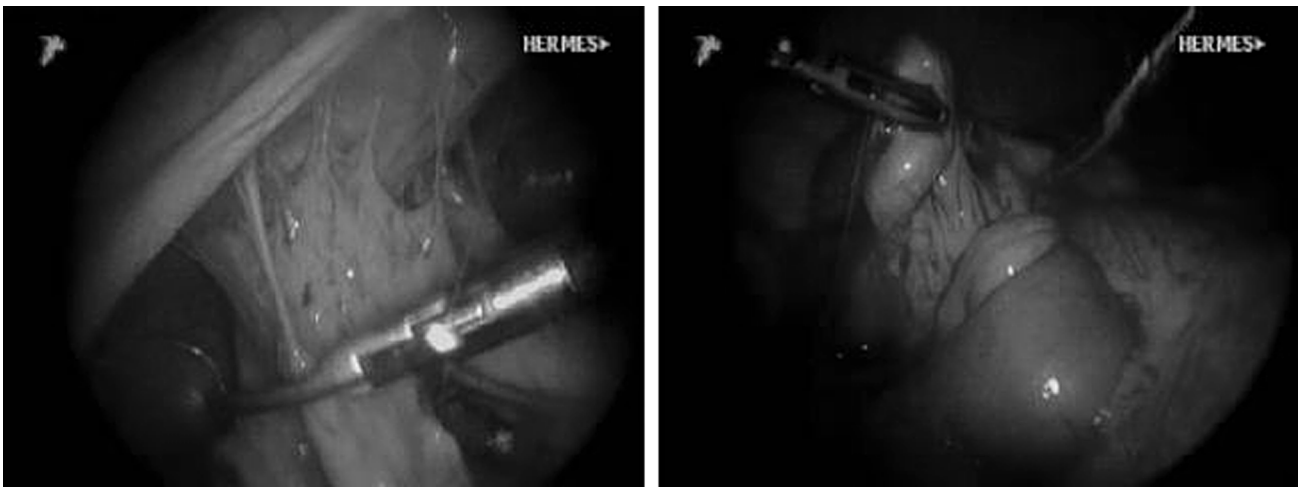

Figure 5. Mild scarring and postinflammatory changes in the right lower quadrant at the time of interval laparoscopic appendectomy 4 months following acute appendicitis, pylephlebitis, and mesenteric venous thrombosis.

Table 1.
Details of the 15 Reported Cases of Mesenteric Venous Thrombosis and Pylephlebitis Complicating Appendicitis in Children Since 1959

Author	Year	Age	Sex	Operative Management	Anticoagulation
Babcock[8]	1979	6	M	Exploratory laparotomy with appendectomy, second laparotomy with liver abscess drainage	No
Shaw[9]	1986	13	M	Exploratory laparotomy with appendectomy and drainage of periappendiceal abscess	No
Giuliano[10]	1989	12	M	Exploratory laparotomy with appendectomy	No
Slovis[11]	1989	6	F	Not stated	Not stated
Slovis[11]	1989	9	M	Not stated	Not stated
Scully (Ed.)[12]	1991	15	M	Exploratory laparotomy with ileocectomy and liver biopsy	No
van Spronsen[13]	1996	11	M	Interval open appendectomy	No
Eire[14]	1998	12	F	Exploratory laparotomy, second-look exploratory laparotomy (occurred subsequent to open appendectomy)	Yes
Kader[15]	1998	15	M	Interval exploratory laparotomy with appendectomy	Yes
Schmutz[16]	1998	18	M	No additional surgery performed (occurred subsequent to appendectomy)	No
Vanamo[17]	2001	7	F	Exploratory laparotomy with appendectomy and drainage periappendiceal abscess	Yes
Chang[18]	2001	8	M	Exploratory laparotomy with ileocectomy and drainage of liver abscesses	Yes
Nanni[19]	2002	10	M	No additional surgery performed (occurred subsequent to appendectomy)	No
Pitcher[20]	2003	17	M	Exploratory laparotomy with appendectomy	Yes
Stitzenberg	2006	5	F	Interval laparoscopic appendectomy	Yes

tients went on to require additional procedures. Two of the 10 patients were treated with interval open procedures, and our current patient was treated with an interval laparoscopic appendectomy. For the remaining 2 patients, no description of the surgical management was included in the report. Of the 3 patients in whom onset of pylephlebitis occurred after appendectomy, 1 patient required subsequent second and third laparotomies, while 2 were managed nonoperatively.

To our knowledge, the case described in this report is the first case of appendicitis complicated by pylephlebitis that was treated using a minimally invasive approach. Using anticoagulation and intravenous antibiotics, we were able to successfully manage the acute presentation nonoperatively. Consequently, we were able to subsequently perform an interval appendectomy laparoscopically. This approach averted the morbidity associated with emergent open procedures.

CONCLUSION

This is the first reported case of minimally invasive management of appendicitis complicated by pylephlebitis and mesenteric venous thrombosis. Based on our experience we feel that, for children with this rare presentation, interval laparoscopic appendectomy, after a course of anticoagulation and intravenous antibiotics, is an acceptable alternative to routine emergent exploratory laparotomy and appendectomy.

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