



# History and Epidemiology of Pediatric Appendicitis

1

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## Introduction

Appendicitis is one of the most common surgical conditions treated in children of all ages. While the appendix was identified and grossly described centuries ago, diseases of the appendix were only recognized a little over 100 years ago. Historic terms such as “perityphlitis” were phased out as we began to understand the pathophysiology and histopathologic changes of acute appendicitis. However, as new definitions emerged, so did new questions. The management of appendicitis in children has been hotly debated since the first surgical therapy was described. Currently, novel operative technology, improved antibiotics, and advanced diagnostic instruments have made their way into the treatment algorithms, shedding insight while also inviting along with them more controversies in the management of pediatric appendicitis.

## The Dark Ages (Pre-Fitz Era)

The appendix was described as early as 1492 by Leonardo da Vinci, though his drawings were not published until several centuries later. Therefore, the Italian anatomist Berengario da Carpi is credited with the first description of the appendix in 1521 as an “empty small cavity at the end of the cecum” [1]. His words were validated by Andreas Vesalius in his illustrations of the colon published in 1543 in *De*

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*Humani Corporis Fabrica*. Shortly thereafter, with a graphic representation available and textual description of the appendix, Gabriel Fallopius compared the appendix to a worm, coining the term “the vermiform appendix” [2]. In the ensuing century, sparse reports of inflammation around the area of the appendix appeared, moving the conversations about the appendix from descriptive anatomy to abnormal findings. In 1711, Heister, an alumnus of Boerhaave, described autopsy findings corresponding to a perforated appendix in the right lower quadrant [2]. Several other authors contributed their postmortem findings in the 1700s such as a blackened appendix, a narrow appendix with abscess, and an obstructed appendix with hardened stool in it. Mestivier described an appendix perforated by a pin and surrounded by “a pint of pus” at the right of the umbilicus [3].

In 1812, John Parkinson presented the case of a 5-year-old with a fecalith leading to perforated appendix with a normal cecum, and in 1813, the first description of pediatric acute appendicitis was presented [4]. Wegeler detailed the clinical presentation and hospital course prior to the demise of an 18-year-old patient with 3 days of abdominal pain, diarrhea, and emesis. Wegeler found a gangrenous cecum with an appendix that was “red, enlarged, and filled with stones” [2]. These two cases marked an important transition where the focus started shifting from postmortem analysis to clinical observations in vivo of the diseases of the appendix. Given this and several other reports of fecal peritonitis, Francois Melier suggested the appendix as the source of the problem and appendectomy as a possible treatment [5]. However, his suggestions fell on deaf ears because the influential Guillaume Dupuytren strongly believed that the inflammatory process began in the cecum and not the appendix. Thanks to Dupuytren, the term perityphlitis continued to be the diagnosis given until the late 1880s [6].

During this time period of discovery, the first appendectomy was performed, though acute appendicitis was not the indication for surgery. In 1735, Claudius Amyand performed the first appendectomy in London. His patient was an 11-year-old boy who was admitted for the repair of a congenital inguinal hernia that had progressed to the point of suppurating a discharge of “an unkindly sort of matter” for 1 month [7]. Amyand found an indirect inguinal hernia containing the appendix, which had been perforated by a pin that the boy swallowed. He describes that “many unsuspected oddities” were found, as his assistants held the boy down during this procedure in the preanesthetic era. Challenging as it was, Amyand’s patient survived the first surgery of the appendix.

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## Renaming and Reframing

While the anatomy of the appendix was recognized early, it had no impact on clinical practice until the 1880s. The modern history of acute appendicitis began in 1886 when Reginald Fitz, pathologist at Harvard, read his paper “Perforating Inflammation of the Vermiform Appendix: With Special Reference to Its Early Diagnosis and Treatment” [8]. This landmark article detailed the presentations of 257 cases of appendicitis, emphasizing that the inflammation in the right lower quadrant, commonly misdiagnosed as perityphlitis, in fact originated from the appendix. In the

same year, Robert Hall performed the first appendectomy for perforated appendicitis in the United States. Three years later, Charles McBurney entered the fray in 1889 and demanded that the “so-called pericecal inflammation” be referred to as appendicitis, functionally removing the term perityphlitis from the medical jargon [9]. McBurney described in detail the constellation of symptoms that we now associate with “classic” appendicitis. He is perhaps better known for his famous depiction of McBurney’s point [9]:

And I believe that in every case the seat of greatest pain, determined by the pressure of one finger, has been very exactly between an inch and a half and two inches from the anterior spinous process of the ilium on a straight line drawn from that process to the umbilicus.

Surgical removal of the appendix became increasingly popular, as surgeons published overwhelmingly positive results with this procedure. McBurney wrote a very detailed case series of 11 patients including their varying clinical presentation and intraoperative findings. He emphasized that the clinical presentation may not match the severity of the disease, and therefore, he firmly recommended immediate operation for all cases. Others, such as Ochsner in 1902, were not as enthusiastic about operating in perforated appendicitis [10]. Ochsner proposed non-operative treatment for peritonitis, with enemas, gastric lavage, and bowel rest, followed by interval appendectomy. This heated debate continued for the better part of the century and, one could argue, still permeates our discussions today. In 1904, John McMurphy added his opinion by reporting his experience with 2000 appendectomies, publishing the largest case series to date and advocating for immediate appendectomy in support of McBurney’s stance, given his low mortality rates [11]. But perhaps the most instrumental event in promoting the surgical treatment of appendicitis was the experience of Sir Frederick Treves. He was summoned by King Edward in 1902 to evaluate him for right lower quadrant pain merely 2 weeks prior to the coronation [12]. The king refused surgical intervention prior to the coronation, which led to a moribund king undergoing abscess drainage weeks later. In the end, he attended the coronation, knowledge of acute appendicitis was publicly disseminated, and appendectomy became the widely accepted treatment.

As a result, the ambition of the era quickly became perfecting surgical technique and mastering the art of surgery. A wide array of surgical approaches were used including transverse laparotomy; midline, paramedian, lateral rectus incisions; oblique incision over the external oblique; and muscle splitting versus cutting incisions [13]. McBurney reported using a right lower quadrant muscle splitting incision. This approach was first used by McArthur, who was unable to present his findings before McBurney. While McBurney admitted this incision was McArthur’s, it was his eponymous name that prevailed. Besides the surgical approach, variations on the technique for the removal of the appendix ranged from simple ligation to purse string on the cecum, crushing at the base with serosal oversewing, or imbrication into the cecum, to name a few [14]. With the vanishing of perityphlitis as a diagnosis, surgeons also encouraged physicians to turn away from dated remedies such as cathartics, which only delayed definitive care. Therefore, the history of appendicitis progressed from naming an unnamed disease to improving treatment and minimizing harm.

A few of the trialed techniques prevailed, while many others faded over time, yet the patient outcomes overall continued to improve. During the following decades, the number of patients diagnosed with acute appendicitis increased exponentially. Initially, as surgical technique was mastered, mortality rates decreased in patients undergoing appendectomy. By the 1930s, overall mortality was as low as 2.1% with lower rates in the uncomplicated cases (0.2–1.0%) and higher in the perforated appendicitis patients (1.6–32%). In McMurphy's personal series, his reported mortality rate decreased from 7% to 2% [11]. However, the total number of deaths per capita from appendicitis remained high, and paradoxically, mortality rates began to rise again to an alarming 10% by 1940. Indignant surgeons attributed such complication rates not to delayed patient presentations, but rather to delayed interventions by surgeons. Morse calls this "procrastination the cause of death – the almost criminal cause" [15]. In the midst of differing opinions on the timing of intervention, pediatric appendicitis began to stand apart from the adult disease. The mortality of appendicitis had risen to 20,000 per year in the United States by 1936 and, in children, had become the 3rd cause of death, only surpassed by trauma and pneumonia in the pre-antibiotic era. Nonetheless, Ladd reported the impeccable mortality record of his own institution with only 2 mortalities in 361 cases [16]. He credited their excellent outcomes to early diagnosis and immediate surgical management. Even Ochsner, from the beginning, had acknowledged children as the one exception to his conservative therapy. The non-operative management of appendicitis in children was not an accepted approach, a claim used to validate the age-old adage that children are not small-sized adults.

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## From Then and to Now

The latter half of the twentieth century was characterized by remarkable advances in the adjunctive modalities for managing acute appendicitis. While the earlier half of the century revolved around perfecting surgical technique, the late 1900s developed improved management algorithms, diagnostic imaging, and advanced surgical instruments. The most notable medical advance of the century was the advent of penicillin in 1928. It wasn't until 1940 when penicillins and sulfonamides were introduced for the treatment of appendicitis with perforation, leading to a dramatic decrease in mortality from 5% to 1.2% [17, 18]. The choice, role, and duration of antibiotic therapy have been variable since then and continue to be an actively studied subject in present-day medicine.

As common themes were recognized, pediatric surgeons began to advocate for protocols and streamlined management of patients presenting with symptoms of appendicitis, in order to intervene prior to the development of peritonitis or perforation. Ravitch stated our goals clearly: "What we seek is a series of criteria that will represent the minimal requirements for operation [to] be safeguarded from ever missing a case [...] and] will not be accumulating a large specimen collection of normal appendices" [19]. To this end, Robert Gross published a detailed, almost prescriptive report of preoperative preparations for appendectomy, including multiple principles to which we still adhere today. He even included his opinion of how long of a delay to surgical therapy was acceptable in children (less than 12 hours) [20].

The 1980s marked another leap in technological advances. The surgical approach to appendectomy had been fairly uniform until 1982, when the gynecologist Kurt Semm introduced laparoscopy as a novel approach [21]. Laparoscopy became quickly accepted in the surgical community, though the techniques used vary greatly between surgeons and institutions. In 1986, ultrasonography became the first noninvasive method for establishing a diagnosis of appendicitis with high accuracy [22]. Computerized tomography did not lag far behind, as the field of radiology was expanding. Most recently, magnetic resonance imaging has emerged as a cross-sectional imaging modality without the drawback of ionizing radiation, and this will be discussed further in later chapter in this book. However, the availability of these technologies and the expertise to operate them are not uniformly available, and, therefore, the optimal imaging strategy is still debated among providers.

Despite the questions that remain unanswered, the outcomes have trended toward improvement since 1886. The annual incidence of pediatric appendicitis is 83 patients per 100,000. The incidence of appendicitis in North America has been rising by 4% per year. The non-perforated appendicitis cases are increasing faster than the perforated ones, which is presumably due to the use of improved imaging modalities and earlier diagnosis [19, 23]. In fact, perforation rates have significantly decreased over time from 50% in 1980 to 20% in 2000 [24]. In the United States, there are 270,000 annual cases of acute appendicitis with the teenagers having the highest incidence [25]. Males have a slightly higher lifetime risk than females (8.6% vs 6.7%) [26]. The mortality in appendectomies has been steadily decreasing from 22% in 1899 down to 0.8% in 1950 and 0.27% by the 1980s [27, 28]. In 2010, there were only 17 deaths per year, according to the census [29]. The mortality is much lower in early appendicitis cases compared to perforated appendicitis with diffuse peritonitis.

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## Conclusion

The optimal management of appendicitis is not always straightforward and has been controversial since its recognition. We have come a long way from the dismissive attitude toward perityphlitis to single-port, laparoscopic appendectomy plus every diagnostic step along the way. Some of our current discrepancies in opinions are rooted in the struggles shared by our predecessors as well. And while our awareness of appendicitis in children is relatively young, surgeons have significantly altered the natural course of the disease. Patient outcomes have become significantly better over time, but there remains room for improvement. This drive to optimize the outcomes for our patients inspires us to address controversies in the field and challenge existing dogma. The following chapters will attempt to address the leading areas of controversy in this field.

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