

Fellow's Column: Spontaneous Intestinal Perforation or Necrotizing Enterocolitis

Kristie Searcy, MD, Shabih Manzar, MD

Summary:

Confusion and overlap exist with the definition of necrotizing enterocolitis (NEC) and spontaneous intestinal perforation (SIP). Although clinically significant NEC and SIP may present similarly, the confirmation requires exploratory laparotomy. The article describes a working algorithm to differentiate between the SIP and NEC. This draft aims to stimulate the neonatology practicing community about this topic. Further discussion is needed to reach some consensus.

Abbreviations:

SIP/FIP- Spontaneous/Focal Intestinal Perforation, NEC- Necrotizing Enterocolitis

“The article describes a working algorithm to differentiate between the SIP and NEC. This draft aims to stimulate the neonatology practicing community about this topic. Further discussion is needed to reach some consensus.”

Cost:

The estimated annual cost of caring for infants with Necrotizing Enterocolitis (NEC) is about \$500 million to \$1 billion. ¹ NEC is used as one of the NICU's key performance indicators. (2,3) A high-performer institution is the one that has the lowest incidence of NEC. However, the data could be influenced by how the institution defines NEC.

“Recently, Nue (4) questioned Bell's criteria for diagnosing NEC, which was developed in the late 1970s. The author believed this staging system is outdated and suggested using the terms' medical NEC' or 'surgical NEC' depending on the clinical symptoms, radiologic signs, and surgical findings.”

Controversy:

Recently, Nue (4) questioned Bell's criteria for diagnosing NEC, which was developed in the late 1970s. The author believed this staging system is outdated and suggested using the terms' medi-

cal NEC' or 'surgical NEC' depending on the clinical symptoms, radiologic signs, and surgical findings. Similarly, Berrington and Embleton (5) expressed concerns about differentiating NEC with focal intestinal perforation (FIP). Swanson et al. (6) reported the published data from a US national data set from 2002 to 2017, showing a decreasing trend in NEC while an increasing trend of spontaneous intestinal perforation (SIP). They also encountered and acknowledged the overlap and misdiagnosis between the two. In a Canadian study, Shah et al. (7) studied a cohort study of 17,426 infants and found higher odds of a composite outcome of mortality or morbidity with NEC than with SIP. In their report, the diagnosis of FIP/SIP was made according to the local practices based on the lack of clinical features of NEC. Studies from Fisher et al. (8) showed that neonates with laparotomy-confirmed SIP had significantly lower mortality than those with laparotomy-confirmed NEC.

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Diagnostic issues:

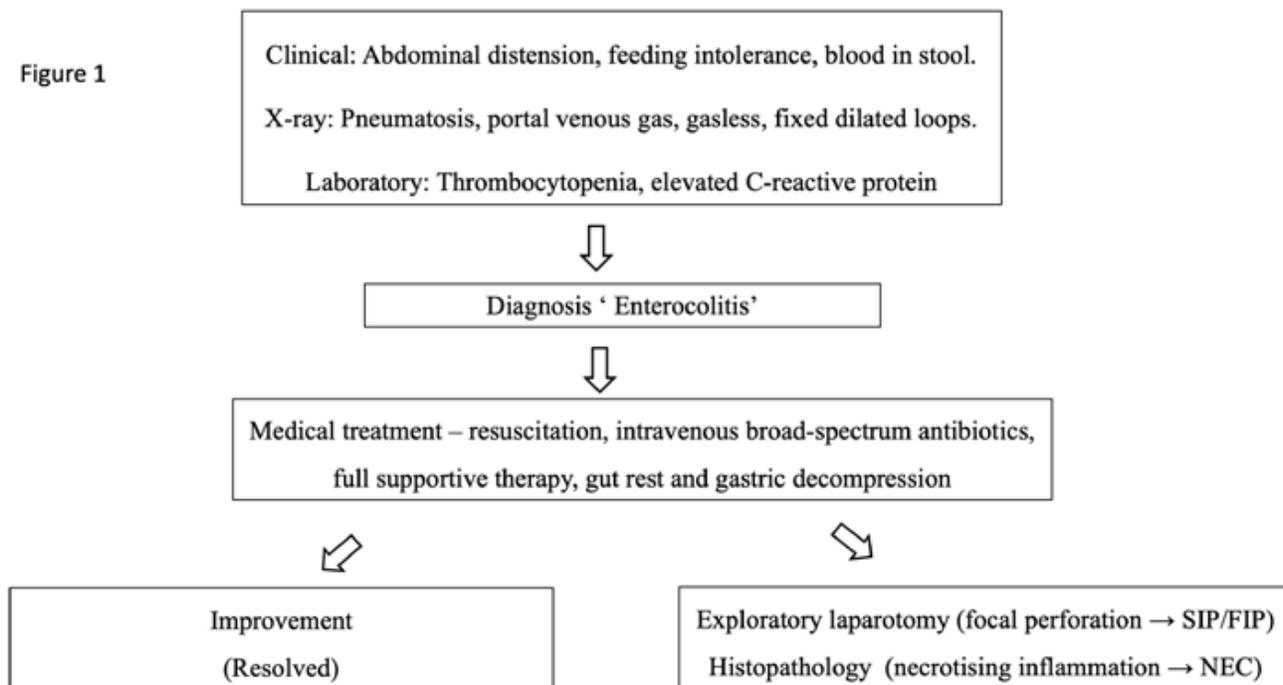
Diagnostic-related groups (DRGs) have been used to measure the complexity of inpatients using a case mix index (CMI). (9) Defining diseases is challenging in neonatal medicine. For example, hyaline membrane disease is also labeled as respiratory distress syndrome. One is a pathological diagnosis, while the other is a clinical one. To diagnose diseases, clinicians rely on clinical presentation supported by radiological, laboratory, and pathological findings. A retrospective case-control study of 114 infants where 48 infants had SPI and 66 had NEC showed that a transient increase in serum alkaline phosphatase level is independently associated with SIP compared to NEC, suggesting utility in using serum alkaline phosphatase levels to differential infants with these conditions. (10)

Labeling a condition with a diagnosis is required for management, data monitoring, and billing. NEC impacts the neurodevelopmental outcome and may persist long term; therefore, data on NEC is essential. For instance, studies suggest that preterm infants diagnosed with surgical NEC had slower catchup head growth at four months and were more likely to experience developmental delay at 36 months compared to preterm infants with surgical SIP. (11) The question is: why do we not create a single clinical diagnosis for conditions with similar clinical presentation. Once the clinician has all the data and evidence, the final diagnosis could be labeled as NEC, SIP/FIP.

Proposed solution:

For example, high bilirubin is entered into the medical record as hyperbilirubinemia. Some clinicians prefer jaundice. Once the fractionated bilirubin information is available, the diagnosis

Figure 1



*Spontaneous Intestinal Perforation, FIP – Focal Intestinal Perforation, NEC –Necrotizing Enterocolitis

Figure 1: The diagnostic flow chart for diagnosis of Enterocolitis to SIP/FIP-NEC

is modified to unconjugated (indirect) or conjugated (direct) hyperbilirubinemia. Further, it could be classified as physiological or pathological jaundice. Using the same analogy, the name for

a clinical presentation for SIP/FIP and NEC could be a common nomenclature.

To diagnose NEC or SIP/FIP, clinicians should look at clinical, ra-

Name	ICD-10 Codes
Enterocolitis	K52.9
Enterococcal bacteremia	R78.81, B95.2
Enterococcal infection	A49.1
Enterococcal sepsis	A41.81
Enterococcal septicemia	A41.81
Enterococcus as the cause of diseases classified elsewhere	B95.2
Enterococcus faecalis infection	B95.2
Enterococcus infection in shunt	T85.78XA, B95.2
Enterococcus UTI	N39.0, B95.2
Enterococcus, vancomycin-resistant	A49.1, Z16.21
Enterococcal fistula	K63.2
Enterocolitis due to Clostridioides difficile	A04.72
Enterocolitis due to Clostridium difficile	A04.72
Enterocolitis due to Clostridium difficile, not specified as recurrent	A04.72
Enterocolitis due to Clostridium difficile, recurrent	A04.71
Enterocolitis of newborn	P78.89, K52.9
Enterocolitis, necrotizing	K55.30
Enterocolitis, necrotizing neonatal	P77.9
Enterocolitis, necrotizing neonatal, stage I	P77.1
Enterocolitis, necrotizing neonatal, stage II	P77.2
Enterocolitis, necrotizing neonatal, stage III	P77.3

Figure 2

Step 1: Select 'Enterocolitis' from the drop-down menu

Step 2: Under modifier free text 'rule of NEC/SIP'

Step 3: Condition improved → Resolved

Step 4: Diagnosis confirmed → NEC or SIP

HCC Weight	Name	ICD-10 Codes	ICD-9 Co
219	Intestinal perforation	K61.1	568.33
	Intestinal perforation	K61.1	568.33
	Incompetent perforator vein	K63.90	454.9
	Incompetent perforator vein, unspecified laterality	K63.90	454.9
	Intestinal perforation, in newborn	P78.0	777.8
	Intestinal perforation, perinate	P78.0	777.8

HCC Weight	Name	ICD-10 Codes
	Necrotizing enterocolitis in newborn, stage I	P77.1
	Necrotizing enterocolitis in newborn, stage II	P77.2
	Necrotizing enterocolitis in newborn, stage III	P77.3

Screen Shots from <https://www.epic.com>

Figure 2: Steps of working on the electronic chart with a diagnosis of Enterocolitis to SIP/FIP-NEC

diological, and laboratory evidence (abdominal distension, feeding intolerance, blood in stool, pneumatosis, portal venous gas, gasless, fixed dilated loops, abnormal laboratory indices).

“Based on this information, a diagnosis of ‘enterocolitis’ could be entertained as, at this point, SIP/NEC could not be ruled out. The caveat with the diagnosis of SIP/FIP and NEC is the need for exploratory laparotomy. Perforation can only be confirmed after surgical intervention, and necrotizing is a histopathological diagnosis.”

Based on this information, a diagnosis of ‘enterocolitis’ could be entertained as, at this point, SIP/NEC could not be ruled out. The caveat with the diagnosis of SIP/FIP and NEC is the need for exploratory laparotomy. Perforation can only be confirmed after surgical intervention, and necrotizing is a histopathological diagnosis. Therefore, starting with a diagnosis of enterocolitis could be justified. Once confirmation is done, the final diagnosis could be changed to FIP or NEC (Figures 1 and 2). As the medical treatment for both conditions is the same (resuscitation, intravenous broad-spectrum antibiotics, full supportive therapy, gut rest, and gastric decompression), differentiating both at the time of occurrence is not that important.

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Kristie Searcy, MD
Internal Medicine-Pediatrics Resident, Department of Pediatrics
LSU Health Sciences Center
1501 Kings Highway
Shreveport, LA 71103

Corresponding Author



Shabih Manzar, MD
Clinical Associate Professor
LSU Health Sciences Center
1501 Kings Highway
Shreveport, LA 71103
Telephone: 318-626-1623
Fax: 318-675-6059
Email: shabih.manzar@lsuhs.edu

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