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Telling the whole COVID-19 story in ICD-10-CM: Capturing the inflammatory response

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As we enter year two of the pandemic in the United States, our knowledge of COVID-19 and its effect on the body continues to evolve. It is clear there are a wide range of presentations in COVID-19 patients, and the focus of this article will be on the most extreme inflammatory manifestations.

First, we need to understand the inflammatory response. Cytokines are molecules released by the immune system in response to infection or other outside insult such as trauma or burns. They circulate in the bloodstream like messengers calling upon other immune cells to fight the offender and assist in the healing process. This is the immune system working correctly.

However, our immune system sometimes acts inappropriately and attacks healthy tissues, resulting in a double insult. Several studies regarding cytokines, published well before the pandemic, have described this type of response as a "cytokine storm," where specific types of cytokines (IL-interleukins) IL-1, IL-6, IL-12, and IL-18 are released, along with tumor necrosis factor alpha (TNFα) and other inflammatory mediators.

This sustained and excessive release continues even after the initial inciting event has resolved, leading to transient or permanent end-organ damage. It can be the cause of death in a variety of infectious diseases including the flu, COVID-19, Ebola and sepsis.

In the extremely ill COVID-19 population, this process has been described as a cytokine storm. Symptoms of a cytokine storm include:

- Fever
- Headache
- Hypotension
- Nausea
- Tachycardia
- Rash
- Respiratory distress

How should the inflammatory presentation be represented in documentation and subsequent coded data? The final answer to this question will certainly evolve as the science evolves.

For COVID-19 patients presenting at the extreme end of the clinical spectrum, potential diagnoses that may be documented include "cytokine storm," CRS, sepsis, multisystem inflammatory syndrome and rarely hemophagocytic lymphohistocystosis (HLH).

CRS

Let's first consider CRS. It was originally identified in patients undergoing CAR-T therapy for cancer and in bone marrow transplants and utilizes a grading system to define the severity of CRS. Grading codes were incorporated into ICD-10-CM on October 1, 2020. Oncologists have developed several grading scales, one of which is described in the Common Terminology Criteria for Adverse Events (CTCAE) system.

CTCAE classifications, which are modified from the National Cancer Institute Common Terminology Criteria for Adverse Events, grade CRS with the corresponding ICD-10-CM code:

- Grade 1: Mild: Asymptomatic or mild symptoms, fever, with or without constitutional symptoms; clinical or diagnostic observations only (D89.831)
- Grade 2: Moderate: Hypotension responding to fluids; hypoxia responding to less than 40% oxygen; Minimal, local or noninvasive intervention indicated (D89.832)
- Grade 3: Severe or medically significant but not immediately life-threatening, and includes ox

requirement of at least 40%, hypotension managed with one vasopressor, hypoxia requiring greater than 40% oxygen (D89.833)
- Grade 4: Life-threatening consequences with urgent intervention indicated. Includes requirement for ventilator support or grade 4 organ toxicity excluding transaminitis (D89. 834)
- Grade 5: Death: Self-explanatory (D89.835)

The specific code assignment will depend on explicit provider documentation of the grade. If no grade is documented, then assign a code for the unspecified grade (D89.839).

Is CRS and the Cancer Institute grading system a "good fit" for COVID-19 patients?

COVID-19 patients can have lab evidence of an exuberant inflammatory response with persistent fevers and elevated proinflammatory cytokines associated with critical and fatal illness; however, according to research, the levels are substantially lower than those seen with CRS. There is also a concern about data integrity if the codes are utilized for conditions outside of the oncology community.

Sepsis
Some would argue that CRS is integral to a diagnosis of sepsis and that in the COVID-19 population "all" patients with CRS would also have sepsis. Varied definitions of sepsis exist in the industry and we recommend organizations follow their own guidelines.

Documentation and coding of sepsis is challenging in that it only "fits" during the immediate infection window. Over the past year, it has become apparent that in some patients with COVID-19, there is a delayed cytokine response after the active COVID-19 infection has peaked.

Labeling a patient without an infectious condition as septic seems inappropriate. So careful review for current infection and sepsis criteria is recommended instead of a blanket approach in obtaining a diagnosis of sepsis on all COVID-19 patients.

Also note that it would be inappropriate to code for systemic inflammatory response syndrome (SIRS) due to non-infectious process during the initial COVID-19 infection because COVID-19 is a viral infection.

There are two remaining conditions that should be evaluated when reviewing a critically ill COVID-19 patient. This is not to suggest the conditions are used for every COVID-19 patient or as a replacement for CRS or sepsis, but as alternative diagnoses providers may want to consider.

Hemophagocytic lymphohistocystosis
Hemophagocytic lymphohistocystosis (HLH; ICD-10-CM code D76.1) is a life-threatening syndrome of excessive immune activation. A viral infection is a common trigger in adults. It is similar to CRS, and in addition to multisystem organ failure, symptoms include:

- Fever
- Cytopenia
- Hepatosplenomegaly
- High serum ferritin
- Neurologic symptoms
- Pulmonary involvement
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In reviewing many recent severe COVID-19 cases, this presentation looks familiar. HLH is uncommon, so unfortunately, providers may not be familiar with HLH and its presentation.

Multisystem inflammatory syndrome (MIS)
Multisystem inflammatory syndrome (MIS; M35.81) includes a child and adult variant (MIS-C and MIS-A). The CDC has posted a well-defined set of clinical criteria for MIS-C, and in October of 2020 they published adult criteria based on several case reports identifying MIS-A in adults, which usually requires intensive care and can have fatal outcomes. The CDC excluded patients if alternative diagnoses such as bacterial sepsis were identified.

Five criteria for the adult population:
1) Severe illness requiring hospitalization in a person 21 years or older.
2) Positive test result for current or previous SARS-CoV-2 infection (nucleic acid, antigen, or antibody) during admission or in the previous 12 weeks.
3) Severe dysfunction of one or more extrapulmonary organ systems (e.g., hypotension or shock, cardiac dysfunction, arterial or venous thrombosis or thromboembolism, or acute liver injury).

4) Laboratory evidence of severe inflammation (e.g., elevated CRP, ferritin, D-dimer, or interleukin-6).
5) Absence of severe respiratory illness (to exclude patients in which inflammation and organ dysfunction might be attributable simply to tissue hypoxia). The CDC included patients with mild respiratory symptoms who otherwise met the criteria.

Five criteria for the pediatric population:

1) Severe illness requiring hospitalization in a person younger than 21
2) Positive for current or recent SARS-CoV-2 infection by RT-PCR, serology, or antigen test or exposure to a suspected or confirmed COVID-19 case within the four weeks prior to the onset of symptoms
3) Evidence of multisystem (more than two) organ involvement (cardiac, renal, respiratory, hematologic, gastrointestinal, dermatologic or neurological)
4) Laboratory evidence of inflammation including, but not limited to, one or more of the following: an elevated C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), fibrinogen, procalcitonin, d-dimer, ferritin, lactic acid dehydrogenase (LDH), or interleukin 6 (IL-6), elevated neutrophils, reduced lymphocytes, and low albumin
5) No alternative plausible diagnoses
6)

The *2021 ICD-10-CM Official Guidelines for Coding and Reporting* were updated January 1, 2021 to include information on the new codes created for COVID-19. Specifically, a new guideline was created on Multisystem Inflammatory Syndrome to provide guidance in sequencing the condition when related to COVID-19 (I.C.1.g.1.l):

- When patients are admitted with both MIS and COVID-19, sequence the COVID-19 (U07.1) as principal diagnosis with MIS (M35.81) as a secondary diagnosis
- When MIS develops because of a previous COVID-19 infection, sequence MIS (M35.81) as principal diagnosis with sequelae of COVID-19 (B94.8) as a secondary diagnosis
- When a patient with history of COVID-19 develops MIS with no provider documentation that the MIS is due to the previous COVID-19 infection, sequence MIS (M35.81) as principal diagnosis with personal history of COVID-19 (Z86.16) as a secondary diagnosis
- When a patient develops MIS with a known or suspected exposure to COVID-19 and no current COVID-19 infection or history of COVID-19, sequence MIS (M35.81) as principal diagnosis with contact with/exposure to COVID-19 (Z20.822) as a secondary diagnosis

To summarize, our knowledge of COVID-19 and its devastating effects is evolving. It is essential we fully represent the clinical picture in documentation and coding so researchers have the critical information needed to investigate the best care options for this population. As our understanding further develops, collaborate with your local experts and providers to determine the appropriate criteria and diagnoses that are appropriate for this population.

Stay safe and wash your hands!

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