

## **Hemolytic Uremic Syndrome (HUS) v1** **Definition Factsheet & Technical Brief**

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meningi^,OR,^neisseria meningi^,OR,^ n meningi^,OR,^
meningococc^,ANDNOT,(^;z23^,OR,^va[cx]^,OR,^shot^,OR,^immun^,OR,^dap^,OR,^dose^,OR,^boost
er^,)
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**Definition Purpose:** Created by the CDC National Syndromic Surveillance Program (NSSP) for use with ED data as part of the rapid reportables project, which seeks to create queries for all National Notifiable Diseases defined by the National Notifiable Diseases Surveillance System (NNDSS). This query is intended to be a version 1 and may be further improved upon by the NSSP community of practice in the future. This query is intended to be narrow and capture visits with specific terms to support greater monitoring and actionable public health data. More complex and/or broader queries may exist that more broadly capture symptoms or travel-based terms.

**New or Revised Definition:** New Definition

**Date Added to ESSENCE:** July 2024

**Definition Description:** Designed to capture emergency department visits associated with post-diarrheal hemolytic uremic syndrome.

**Use-Cases for Definition:**

- Case-finding
- Trend monitoring
- Early outbreak detection

**Justification:** No query definition exists for post-diarrheal hemolytic uremic syndrome (HUS) infections. CDC wants to use syndromic data to monitor trends.

**Data Source During Development:** Emergency Department Visits. Chief Complaint Query Validation dataset (CCQV) in ESSENCE.

**Development Methods:**

1. The CDC team used the NNDSS case definition of Hemolytic Uremic Syndrome (HUS) to identify diagnostic codes (International Classification of Diseases, 9th & 10th Revisions (ICD-9-CM, ICD-10-CM), SNOMED Clinical Terms (CT)) and chief complaint free text for syndrome development.
2. The codes and free text were then categorized as inclusion or exclusion terms. These terms were formatted for querying in ESSENCE.
3. Text analysis was conducted on the initial query to find additional inclusion or exclusion terms.
4. The query was cross-checked internally by NSSP and by CDC subject matter experts as needed.

Validation Methods: This definition is part of the collection of definitions created by CDC NSSP for the rapid reportables project. The rapid reportables project is led by the CDC NSSP team to create definitions for all nationally reportable diseases. These definitions focus on chief complaint text and diagnostic codes of the disease or its causative agents with no mention of symptoms. Select members of the Community of Practice were then asked to review line-level results for their jurisdictions and provide feedback to ensure the intended results of the definitions were met.

Definition Fields and Structure:

The Chief Complaint Discharge Diagnosis (CCDD) parsed field is used to query the discharge diagnosis codes and chief complaint free text.

- Inclusion based on HUS-related chief complaint free-text terms:
  - The visit is included if it contains:
    1. Chief complaints related to the disease name – (“hemolytic uremic syndrome”, “hus”) **OR**
- Inclusion based on ICD-10 discharge diagnosis codes related to HUS:
  - The visit is included if it contains:
    1. A discharge diagnosis code of Hemolytic-uremic syndrome, unspecified (D59.30) **OR**
    2. A discharge diagnosis code of Infection-associated hemolytic-uremic syndrome (D59.31) **OR**
    3. A discharge diagnosis code of Other hemolytic-uremic syndrome (D59.39) **OR**
- Inclusion based on SNOMED CT discharge diagnosis codes related to HUS:
  - The visit is included if it contains:
    1. A discharge diagnosis code of Hemolytic uremic syndrome (111407006) **OR**
    2. A discharge diagnosis code of Diarrhea-associated hemolytic uremic syndrome (373421000) **OR**
    3. A discharge diagnosis code of Atypical hemolytic uremic syndrome (789660001) **OR**
    4. A discharge diagnosis code of Hemolytic uremic syndrome, adult type (78209002) **OR**
    5. A discharge diagnosis code of Hemolytic uremic syndrome of childhood (36568005) **OR**
    6. A discharge diagnosis code of Infection-related hemolytic uremic syndrome (1269270002) **AND**
- The above inclusion terms must also include terms related to gastrointestinal (GI) organisms that are typically associated with post-diarrheal HUS:
  - Chief complaint free-text terms related to diarrhea and GI organisms such as *E. coli* – (“diarrhea”, “*e coli*”, “*ecoli*”, “shiga toxin”, “STEC”, “*shigella*”) **OR**
  - ICD-10 discharge diagnosis codes related to diarrhea and GI organisms:
    1. A discharge diagnosis code of Diarrhea (R19.7) **OR**
    2. A discharge diagnosis code of *Escherichia coli* [*E. coli*] as the cause of the disease classified elsewhere (B96.2) **OR**
    3. A discharge diagnosis code of Sepsis due to *Escherichia coli* [*E. coli*] (A41.51) **OR**
    4. A discharge diagnosis code of Contact with and (suspected) exposure to intestinal infectious diseases due to *Escherichia coli* (*E. coli*) (Z20.021) **OR**
    5. A discharge diagnosis code of *Shigella* due to Shigellosis due to *Shigella dysenteriae* (A03.0) **OR**
  - SNOMED discharge diagnosis codes related to diarrhea and GI organisms:
    1. A discharge diagnosis code of Acute diarrhea (409966000) **OR**
    2. A discharge diagnosis code of Infection caused by *Escherichia coli* (71057007) **OR**
    3. A discharge diagnosis code of Intestinal infection caused by *Escherichia coli* (111839008) **OR**
    4. A discharge diagnosis code of Infection caused by Shiga toxin producing *Escherichia coli* (328291000119103) **ANDNOT**
- All of the above must exclude:

- Chief complaint free-text terms related to history of HUS – (“hx of hus”, “history of hus”, “thrombocytopenic purpura”, “TTP”) **OR**
- ICD-10 discharge diagnosis codes related to hereditary HUS:
  1. A discharge diagnosis code of Hereditary hemolytic-uremic syndrome (D59.32) **OR**
- SNOMED discharge diagnosis codes related to non-acute onset HUS:
  1. A discharge diagnosis code of Diarrhea-negative hemolytic uremic syndrome (373422007) **OR**
  2. A discharge diagnosis code of Familial hemolytic uremic syndrome (722721004) **OR**
  3. A discharge diagnosis code of Congenital hemolytic uremic syndrome (444976001) **OR**
  4. A discharge diagnosis code of Hemolytic uremic syndrome with diacylglycerol kinase epsilon deficiency (1269225005) **OR**
  5. A discharge diagnosis code of Atypical hemolytic uremic syndrome with complement gene abnormality (1293264009) **OR**
  6. A discharge diagnosis code of Upshaw-Schulman syndrome (373420004)

Detailed Definition Components

**Table 1.** Inclusion and exclusion terms related to HUS and GI organisms. Inclusion terms related to HUS were combined with terms related to GI organisms that are associated with post-diarrheal HUS.\*

Variable Type	Terms	Description
<b><i>Inclusions</i></b>		
Discharge Diagnosis: ICD-10-CM	D59.30	Hemolytic-uremic syndrome, unspecified
	D59.31	Infection-associated hemolytic-uremic syndrome
	D59.39	Other hemolytic-uremic syndrome
	R19.7*	Diarrhea
	B96.2*	<i>Escherichia coli</i> [ <i>E. coli</i> ] as the cause of disease classified elsewhere
	A41.51*	Sepsis due to <i>Escherichia coli</i> [ <i>E. coli</i> ]
	Z20.021*	Contact with and (suspected) exposure to intestinal infectious diseases due to <i>Escherichia coli</i> ( <i>E. coli</i> )
	A03.0*	Shigellosis due to <i>Shigella dysenteriae</i>
Discharge Diagnosis: SNOMED concepts	111407006	Hemolytic uremic syndrome
	373421000	Diarrhea-associated hemolytic uremic syndrome
	789660001	Atypical hemolytic uremic syndrome
	78209002	Hemolytic uremic syndrome, adult type
	36568005	Hemolytic uremic syndrome of childhood
	1269270002	Infection-related hemolytic uremic syndrome
	409966000*	Acute diarrhea
	71057007*	Infection caused by <i>Escherichia coli</i>
	111839008*	Intestinal infection caused by <i>Escherichia coli</i>
328291000119103*	Infection caused by Shiga toxin producing <i>Escherichia coli</i>	
Chief Complaint Terms	hemolytic uremic syndrome	
	hus	
	diarrhea*	
	<i>e coli</i> *	
	<i>ecoli</i> *	
	shiga toxin*	
	STEC*	
	<i>shigella dysent</i> *	
<i>s dysent</i> *		
<b><i>Exclusions</i></b>		
Discharge Diagnosis: ICD-10-CM	D59.32	Hereditary hemolytic-uremic syndrome
Discharge Diagnosis: SNOMED concepts	373422007	Diarrhea-negative hemolytic uremic syndrome
	722721004	Familial hemolytic uremic syndrome
	444976001	Congenital hemolytic uremic syndrome
	1269225005	Hemolytic uremic syndrome with diacylglycerol kinase epsilon deficiency
	1293264009	Atypical hemolytic uremic syndrome with complement gene abnormality
373420004	Upshaw-Schulman syndrome	
Chief Complaint Terms	hx of hus	history of hus
	thrombocytopenic purpura	TTP

**Table 2.** *ESSENCE* syntax

(, (^hemolytic uremic syndrome^, or, !hus!, or, ^;D593[019]^, or, ^;111407006^, or, ^;373421000^, or, ^;789660001^, or, ^;78209002^, or, ^;36568005^, or, ^;1269270002^, ^), and, (^diarrhea^, or, ^;R197^, or, ^;409966000^, or, ^e coli^, or, ^ecoli^, or, ^;B962^, or, ^;A4151^, or, ^;Z20021^, or, ^;71057007^, or, ^;111839008^, or, ^shiga toxin^, or, !STEC!, or, ^;328291000119103^, or, ^shigella dysent^, or, ^s dysent^, or, ^;A030^, ^), andnot, (^hx of hus^, or, ^history of hus^, or, ^thrombocytopenic purpura^, or, ^TTP, or, ^;D5932^, or, ^;373422007^, or, ^;722721004^, or, ^;444976001^, or, ^;1269225005^, or, ^;1293264009^, or, ^;373420004^, ^,)