Idiopathic Pericarditis and Pericardial Effusion in Children: Contemporary Epidemiology and Management

Divya Shakti, MBBS, MPH; Rebecca Hehn, MA; Kimberly Gauvreau, ScD; Robert P. Sundel, MD; Jane W. Newburger, MD, MPH

**Background**—Multicenter studies on idiopathic or viral pericarditis and pericardial effusion (PPE) have not been reported in children. Colchicine use for PPE in adults is supported. We explored epidemiology and management for inpatient hospitalizations for PPE in US children and risk factors for readmission.

**Methods and Results**—We analyzed patients in the Pediatric Health Information System database for (1) a code for PPE; (2) absence of codes for underlying systemic disease (eg, neoplastic, cardiac, rheumatologic, renal); (3) age ≥30 days and <21 years; and (4) discharge between January 1, 2007, and December 31, 2012, from 38 hospitals contributing complete data for each year of the study period. Among 11 364 hospitalizations with PPE codes during the study period, 543 (4.8%) met entry criteria for idiopathic or viral PPE. Significantly more boys were noted, especially among adolescents. No temporal trends were noted. Median age was 14.5 years (interquartile range 7.3 to 16.6 years); 78 patients (14.4%) underwent pericardiocentesis, 13 (2.4%) underwent pericardiectomy, and 11 (2.0%) underwent pericardiectomy; 157 (28.9%) had an intensive care unit stay, including 2.0% with tamponade. Median hospitalization was 3 days (interquartile range 2 to 4 days). Medications used at initial admission were nonsteroidal anti-inflammatory drugs (71.3%), corticosteroids (22.7%), aspirin (7.0%), and colchicine (3.9%). Readmissions within 1 year of initial admission occurred in 46 of 447 patients (10.3%), mostly in the first 3 months. No independent predictors of readmission were noted, but our statistical power was limited. Practice variation was noted in medical management and pericardiocentesis.

**Conclusions**—Our report provides the first large multicenter description of idiopathic or viral PPE in children. Idiopathic or viral PPE is most common in male adolescents and is treated infrequently with colchicine. *(J Am Heart Assoc. 2014;3:e001483 doi: 10.1161/JAHA.114.001483)*

**Key Words:** colchicine • pediatric • pericardial disease • pericardial effusion • pericarditis • pericardium

Idiopathic pericarditis accounts for 37% to 68% of inpatient admissions for pericardial effusions or acute pericarditis in children.1–3 The usual approach to initial management of acute pericarditis includes administration of nonsteroidal anti-inflammatory agents. Colchicine has emerged as an important adjunct in management of recurrent pericarditis and acute pericarditis in adults.4,5 The European Society of Cardiology guidelines for management of pericarditis recommend use of colchicine in recurrent and acute pericarditis6; however, management may include other pharmacological agents, and invasive procedures for pericardial effusions are sometimes needed. Recurrence of pericarditis is common in both children and adults, with reports ranging from 15% to 40%.2–4,7,8 Higher recurrence rates have been observed in patients with a history of use of corticosteroids in the initial episode of pericarditis in adults.4,7 Data on epidemiology, management guidelines, and colchicine use in children for pericarditis are sparse.9,10

Most studies on pericarditis in children have been single-center series.1,3,9 In the present multicenter study, we sought to characterize epidemiology, associated morbidities, inpatient management, and risk factors for readmission in idiopathic or viral pericarditis and pericardial effusion (PPE). Our analyses were performed using the Pediatric Health Information System (PHIS) database of freestanding children’s hospitals in the United States.

**Methods**

**Study Design**

We performed a retrospective review and analysis of a multicenter database. Approval for the study was obtained...
from the institutional review board at the Boston Children’s Hospital, which waived the informed consent requirement.

Data Source
Data were obtained from the PHIS database for the 6-year period encompassing calendar years 2007–2012. The PHIS database is a resource utilization database from participating freestanding children’s hospitals in the United States. It provides information for all hospital discharges including demographic data; diagnostic and procedural International Classification of Diseases, ninth revision (ICD-9) codes (level 1) and resource utilization data (level 2) from charges for procedures, medications, imaging studies, laboratory tests, supplies, and other clinical services. The PHIS database allows longitudinal follow-up of patients who have multiple admissions at the same hospital. The data quality is ensured through systematic monitoring.

Selection Process
From the PHIS database, we selected all inpatient hospital discharges between January 1, 2007, and December 31, 2012, with ICD-9 codes suggesting PPE at a hospital that contributed both level 1 and level 2 data to PHIS for each year of the study period. Inclusion criteria were age <21 years at admission; ICD-9 diagnosis codes indicating PPE or procedure codes indicating pericardiocentesis, pericardiectomy, or pericardiotomy (see Table S1); and no prior admission with pericarditis codes in the preceding year.

Etiologies for PPE
We identified patients with underlying systemic diseases, neonatal problems, and cardiac surgery as having potential causes for PPE and assigned the following appropriate etiologic categories (see Tables S2 and S3):
1. ICD-9 codes for underlying systemic diseases or other potential causes for PPE (e.g., neoplastic, cardiac, myocarditis, endocarditis, rheumatic, rheumatologic, chronic renal, endocrine, immunologic, chest trauma, organ transplant, nonviral infection, hematologic, gastrointestinal, hematologic, and perinatal) were used to generate categories for possible etiology of pericarditis.
2. Underlying neonatal problems such as birth asphyxia, hydrops fetalis, prematurity (<36 weeks of gestation), low birth weight (<2500 g), and patients aged <31 days were noted.
3. Repair of congenital heart defect in the past 6 months, as defined by the entry criteria for the Risk Adjusted classification for Congenital Heart Surgery (RACHS-1) method, and placement of a pacemaker (epicardial and transvenous) were noted.11

From the cohort of patients with PPE, we identified patients with likely idiopathic or viral PPE by excluding all those with other potential causes for PPE (see Table S4).

Data Obtained
Variables collected from the PHIS database were patient age, sex, birth weight, center of care, race, geographic region, dates of admission and discharge, stay in the intensive care unit, mortality, principal and secondary diagnosis codes, procedure codes, medications (anti-inflammatory medications and antacids). The study period was divided by year and by quarter to assess uniformity in the number of pericarditis discharges over time.

Initial Admission
Initial admission was defined as admission with qualifying diagnosis or procedure codes for PPE if the patient was not admitted with qualifying diagnosis or procedure codes in the preceding year (including up to 2006).

Readmission
Readmission was defined as subsequent admission with qualifying diagnosis or procedure codes for PPE within 1 year of initial admission. We limited our sample to those patients whose initial admission was between 2007 and 2011, allowing for a complete year of follow-up. Readmissions at the same center with PPE were identified using unique patient identifiers in the PHIS database.

Statistical Analysis
Patient and procedural characteristics were summarized using frequencies and percentages for categorical variables and medians with 25th and 75th percentiles (interquartile range) for continuous variables. The chi-square goodness of fit test was used to explore uniformity with respect to the number of discharges throughout time (quarterly and yearly). Logistic regression was used to assess relationships between patient factors and the outcome of readmission within 12 months. Odds ratios with 95% confidence intervals are reported. Variables significant at the 0.10 level were considered for inclusion in a multivariable model. P≤0.05 was considered significant. SAS v9.3 (SAS Institute, Cary NC) was used for data analysis.

Results
A total of 11 364 eligible patients were noted in the 38 centers that contributed both level 1 and level 2 data to
the PHIS during each of the 6 years studied. Of these, we excluded 10,802 patients who had codes for diseases known to be associated with pericarditis (see Table S4) and 19 patients aged <31 days. The remaining 543 patients were presumed to have idiopathic or viral PPE and constituted the cohort for our study. The demographic and clinical characteristics of our study cohort are shown in Table 1.

Subjects’ median age was 14.5 years (interquartile range 7.3 to 16.6 years). In the adolescent age group, idiopathic or viral PPE was significantly more frequent in boys than in girls (Figure 1), and male adolescents constituted about half of our study cohort. The Northeast region had a significantly higher number of hospital discharges with idiopathic or viral PPE per 100,000 hospital discharges ($P<0.001$) (Figure 2); however, we found no significant quarterly or yearly association in the number of pericarditis discharges during the study period (Figure 3).

**Hospital Course**

There were no inpatient deaths in our cohort. The median length of hospital stay was 3 days (interquartile range 2 to 4 days). Care in an intensive care unit was noted for 157 patients (28.9%), and cardiac tamponade was coded for 11 (2.0%). A pericardial drainage procedure was performed in 96 patients (17.7%), including (not mutually exclusive) pericardiocentesis in 78 (14.4%), pericardiotomy in 13 (2.4%), and pericardiectomy in 11 (2.0%).

**Table 1. Demographic and Clinical Characteristics of Patients With Idiopathic or Viral Pericarditis and Pericardial Effusion**

<table>
<thead>
<tr>
<th>Characteristic (N=543)</th>
<th>Median (IQR) or n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Demographic</td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>157 (28.9)</td>
</tr>
<tr>
<td>Age, y</td>
<td>14.5 (7.3 to 16.6)</td>
</tr>
<tr>
<td>Race</td>
<td></td>
</tr>
<tr>
<td>Black</td>
<td>113 (20.8)</td>
</tr>
<tr>
<td>Hispanic or Latino</td>
<td>75 (13.8)</td>
</tr>
<tr>
<td>White</td>
<td>277 (51.0)</td>
</tr>
<tr>
<td>Other or unknown</td>
<td>78 (14.4)</td>
</tr>
<tr>
<td>Clinical</td>
<td></td>
</tr>
<tr>
<td>Hospital length of stay (days)</td>
<td>3 (2 to 4)</td>
</tr>
<tr>
<td>Intensive care unit stay</td>
<td>157 (28.9)</td>
</tr>
<tr>
<td>Cardiac tamponade</td>
<td>11 (2.0)</td>
</tr>
<tr>
<td>Pericardial drainage procedure*</td>
<td>96 (17.7)</td>
</tr>
</tbody>
</table>

IQR indicates interquartile range.  
*Includes pericardiocentesis, pericardiotomy, and pericardiectomy procedures.

**Medications**

At least 1 anti-inflammatory medication was used in 433 patients (79.7%). Nonsteroidal anti-inflammatory drugs were used most commonly (387 patients, 71.3%), followed by corticosteroids (123 patients, 22.7%), aspirin (38 patients, 7%), colchicine (21 patients, 3.9%), and methotrexate (2 patients, 0.4%). Use of these medications was not mutually exclusive. Other immunosuppressants, such as anakinra, azathioprine, cyclophosphamide, cyclosporine, and rituximab, were not used in initial admissions. Pharmacological management in initial admission is shown in Table 2. More than 1 anti-inflammatory medication was used in 100 patients (18.4%). Aspirin or nonsteroidal anti-inflammatory drugs were used alone in more than half of cases, followed by their combination with corticosteroids (14.6%). Colchicine was always used in combination with other anti-inflammatory medications. These medications could have been used either
Readmissions
Readmission at least once within 1 year of initial admission with ICD-9 codes for PPE occurred in 46 of 447 patients who had at least 1 year of follow-up (10.3%). Most of these patients had only a single readmission (n=33, 71.7%). Multiple...
readmissions within 1 year of initial admission were noted in 13 patients: 2 readmissions were noted for 8 patients, 3 readmissions were noted for 4, and 4 readmissions were noted for 1. Figure 5 shows the time to readmission for these patients. Most (78.3%) were readmitted within 3 months of the initial admission.

Risk factors for readmission are summarized in Table 3. On univariate analysis, geographic region had a borderline association with readmission ($P=0.051$); the highest odds ratio for readmission, compared with the West, occurred in the Midwest region (5.47; 95% CI, 1.53 to 19.5). We did not find significant associations of readmission with other factors, including sex, age, race, intensive care unit admission, hospital lengths of stay, or use of pericardial drainage procedures on first admission. None of the medications used in initial admission was significantly associated with readmission. Multivariable analysis demonstrated that region continued to have a borderline association with readmission ($P=0.05$).

### Discussion

Using the PHIS database from 2007 to 2012, we describe the epidemiology of idiopathic or viral PPE in children requiring inpatient care in freestanding children’s hospitals. Adolescents, particularly boys, were most commonly affected. We found no temporal trends. Although codes for cardiac tamponade were uncommon, pericardial drainage procedures were used in 18% of patients on initial admission. Nonsteroidal anti-inflammatory drugs or aspirin were used most commonly, either alone or in combinations with other medications, followed by corticosteroid use; however, colchicine was uncommonly administered. Practice variation appeared to be marked, although small numbers prevented us from testing statistical significance. Readmission with PPE was noted for 1 in 10 patients, mostly within 3 months of initial admission. Although use of corticosteroids in adults with pericarditis has been associated with a higher complication rate including increased recurrence rate, we could not identify significant risk factors for readmission in our cohort with very limited power. Our data do not allow us to understand the trend toward higher odds of readmission in the Midwest region, although practice variation may be a contributing factor.

Although the incidence of idiopathic or viral PPE appeared to rise during adolescence in both sexes, the spike in male adolescents was especially striking. Although we could not find previous reports of a male predilection for idiopathic or viral PPE, male predominance has been noted in cases of viral myocarditis. Myocarditis and pericarditis may share common viral etiologic agents. Moreover, mouse models of myocarditis have demonstrated the influence of sex-associated steroid hormones on viral concentration, disease susceptibility, and immune response to infection. The interplay of hormonal changes in adolescence and male susceptibility to viral infections is likely to play a role; however, we cannot exclude other etiologic factors for male predominance.

### Table 3. Risk Factors for Readmission on Univariate Analysis

<table>
<thead>
<tr>
<th>Variable</th>
<th>Odds Ratio (95% CI)</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female sex</td>
<td>0.66 (0.32 to 1.37)</td>
<td>0.25</td>
</tr>
<tr>
<td>Region</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Midwest</td>
<td>5.47 (1.53 to 19.49)</td>
<td>0.05</td>
</tr>
<tr>
<td>Northeast</td>
<td>2.70 (0.71 to 10.33)</td>
<td></td>
</tr>
<tr>
<td>South</td>
<td>3.32 (0.95 to 11.62)</td>
<td></td>
</tr>
<tr>
<td>West</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Race</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Black</td>
<td>1.65 (0.78 to 3.46)</td>
<td>0.6</td>
</tr>
<tr>
<td>Hispanic or Latino</td>
<td>1.41 (0.57 to 3.52)</td>
<td></td>
</tr>
<tr>
<td>White</td>
<td>1.0</td>
<td></td>
</tr>
<tr>
<td>Other or unknown</td>
<td>1.13 (0.43 to 2.93)</td>
<td></td>
</tr>
<tr>
<td>1 year increase in age</td>
<td>0.97 (0.93 to 1.02)</td>
<td>0.28</td>
</tr>
<tr>
<td>ICU admission</td>
<td>0.99 (0.50 to 1.95)</td>
<td>0.98</td>
</tr>
<tr>
<td>1 day increase in hospital LOS</td>
<td>0.99 (0.92 to 1.05)</td>
<td>0.67</td>
</tr>
<tr>
<td>Pericardial procedure</td>
<td>1.57 (0.76 to 3.24)</td>
<td>0.23</td>
</tr>
<tr>
<td>Antacid</td>
<td>1.90 (0.96 to 3.78)</td>
<td>0.07</td>
</tr>
<tr>
<td>Corticosteroid</td>
<td>1.56 (0.80 to 3.04)</td>
<td>0.20</td>
</tr>
<tr>
<td>Aspirin</td>
<td>1.22 (0.41 to 3.65)</td>
<td>0.72</td>
</tr>
<tr>
<td>NSAID</td>
<td>1.26 (0.62 to 2.57)</td>
<td>0.52</td>
</tr>
<tr>
<td>Colchicine</td>
<td>1.80 (0.50 to 6.45)</td>
<td>0.37</td>
</tr>
</tbody>
</table>

ICU indicates intensive care unit; LOS, length of stay; NSAID, nonsteroidal anti-inflammatory drug.
The use of colchicine was concentrated in a few pediatric centers; however, colchicine has been shown to reduce recurrence rate, duration of symptoms, and hospitalization rate in multiple adult studies, including a recent randomized double-blind placebo-controlled trial in adults with acute pericarditis.\textsuperscript{4,19} Moreover, addition of colchicine to nonsteroidal anti-inflammatory drugs or aspirin has not been associated with a greater rate of serious adverse side effects.\textsuperscript{4,18,19} Colchicine has been recommended in acute (class 2a) and recurrent (class 1) pericarditis by the European Society of Cardiology since 2004.\textsuperscript{6} These guidelines and adult studies demonstrating benefits of colchicine use without significant adverse effects predate our study cohort. Colchicine has been used in children with familial Mediterranean fever.\textsuperscript{20} Its use has also been reported in small pediatric series of pericarditis.\textsuperscript{9,10} Our data suggest that the practice of colchicine use in adults with pericarditis or PPE has not yet translated to pediatric practice. The infrequent use of colchicine in children with viral or idiopathic PPE might be related to the paucity of published pediatric studies on its use or to a lack of familiarity with this drug in pediatric cardiology practice.

Our study should be interpreted in light of its limitations. Related to the use of administrative databases, limitations include omission of diagnoses or medications because of errors in data entry or coding as well as coding of only limited clinical data. Because children’s weights are not available, we were unable to report doses of medications. Our data derived from a database of pediatric inpatients admitted to freestanding children’s hospitals and may not be generalizable to pediatric admissions in other inpatient settings or to outpatient management. Patients transferred to PHIS hospitals after initial treatment elsewhere may be incorrectly captured as initial cases. The inability to track patients across centers could have contributed to underestimation of readmissions. The recurrence rate of pericarditis could be further underestimated by the absence of data on outpatient management and evaluation in the emergency room. Eleven patients had codes for pericardiectomy on initial admission as defined in our entry criteria, yet pericardiectomy is usually performed in patients with chronic recurrent pericarditis. We are unable to exclude the possibility that this finding is due to coding error or misclassification, for example, because of our inability to track patients across centers. Finally, the small number of readmissions limited our power to find significant predictors of readmission in our study.

In conclusion, we report the first large multicenter description of idiopathic or viral PPE in children and explore the risk factors for readmission. Our data suggest that idiopathic or viral PPE is most common in adolescents, particularly boys, and that practices in the care of these patients vary across centers. Further multicenter studies or a prospective registry would be helpful in formulating recommendations for therapy in children.

Sources of Funding
This work was supported by the Kostin Family Innovation Fund and the Farb Family Fund.

Disclosures
None.

References


Supplementary Material

Supplementary Table 1: ICD-9 Diagnosis and Procedure codes along with their description, used for selection of discharges with pericarditis or pericardial effusion.

<table>
<thead>
<tr>
<th>ICD-9 Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Diagnosis Code</strong></td>
<td></td>
</tr>
<tr>
<td>423.9</td>
<td>Pericarditis (granular) (with decompensation) (with effusion), unspecified disease of pericardium</td>
</tr>
<tr>
<td>420.90</td>
<td>Acute (non-rheumatic) pericarditis , unspecified- hemorrhagic, infective NOS, neoplastic, sicca,</td>
</tr>
<tr>
<td>420.99</td>
<td>Bacterial, fibrinopurulent , fibrinous pericarditis- (acute) (subacute) (with serous or seropurulent effusion), pneumococcal, staphylococcal, streptococcal, suppurative, purulent, septic pericarditis, pyopericardium pneumopyopericardium</td>
</tr>
<tr>
<td>420.91</td>
<td>Benign, idiopathic, non-specific, viral pericarditis</td>
</tr>
<tr>
<td>423.1</td>
<td>Adhesive, adherent, fibrous (external) (internal), obliterans, obliterating, plastic pericarditis, Milk spots, Soldier’s patches.</td>
</tr>
<tr>
<td>420.0</td>
<td>Acute pericarditis in diseases classified elsewhere (coded separately)</td>
</tr>
<tr>
<td>423.2</td>
<td>Calcareous, constrictive pericarditis, Concato’s disease, Pick’s disease of heart (and liver)</td>
</tr>
<tr>
<td>423.3</td>
<td>Cardiac tamponade</td>
</tr>
<tr>
<td>423.8</td>
<td>Chronic (non-rheumatic) neoplasic pericarditis, other unspecified diseases of pericardium</td>
</tr>
<tr>
<td>074.21</td>
<td>Coxsackie pericarditis</td>
</tr>
<tr>
<td>391.0</td>
<td>Acute rheumatic pericarditis</td>
</tr>
<tr>
<td>Code</td>
<td>Description</td>
</tr>
<tr>
<td>-------</td>
<td>-----------------------------------------------------------------------------</td>
</tr>
<tr>
<td>393</td>
<td>Inactive or quiescent (Chronic) Rheumatic – (pericarditis, mediastinopericarditis, myopericarditis)</td>
</tr>
<tr>
<td>115.03</td>
<td>Infection by Histoplasma capsulatum, pericarditis</td>
</tr>
<tr>
<td>115.13</td>
<td>Infection by Histoplasma duboisii, pericarditis</td>
</tr>
<tr>
<td>115.13</td>
<td>Infection by Histoplasma duboisii, pericarditis</td>
</tr>
<tr>
<td>115.93</td>
<td>Histoplasmosis, unspecified pericarditis</td>
</tr>
<tr>
<td>098.83</td>
<td>Gonococcal pericarditis</td>
</tr>
<tr>
<td>423.0</td>
<td>Hemopericardium</td>
</tr>
<tr>
<td>036.41</td>
<td>Meningococcal pericarditis</td>
</tr>
<tr>
<td>093.81</td>
<td>Syphilitic pericarditis</td>
</tr>
<tr>
<td>420</td>
<td>Acute pericarditis (mediastinopericarditis, myopericarditis, pericardial effusion, pleuropericarditis, pneumopericarditis)</td>
</tr>
<tr>
<td>420.9</td>
<td>Other and unspecified acute pericarditis</td>
</tr>
<tr>
<td>411.0</td>
<td>Dressler’s (post myocardial infarction syndrome)</td>
</tr>
</tbody>
</table>

**Procedure Codes**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>37.0</td>
<td>Pericardiocentesis</td>
</tr>
<tr>
<td>37.12</td>
<td>Pericardiotomy</td>
</tr>
<tr>
<td>37.31</td>
<td>Pericardiection</td>
</tr>
</tbody>
</table>
Supplementary Table 2: ICD-9 Codes categorized for possible etiologies for pericarditis

1. **Cardiac Surgery/Procedure**

   - RACHS-1 entry criteria codes

     **Procedure code:**
     - 39.6 Extracorporeal circulation and procedures auxiliary to heart surgery

     **Procedure codes for pacemaker placement:**
     - 37.7 Insertion, revision, replacement, and removal of leads; insertion of temporary pacemaker system; or revision of cardiac device pocket
     - 37.70 Initial insertion of lead [electrode], not otherwise specified
     - 37.71 Initial insertion of transvenous lead [electrode] into ventricle
     - 37.72 Initial insertion of transvenous leads [electrodes] into atrium and ventricle
     - 37.73 Initial insertion of transvenous lead [electrode] into atrium
     - 37.75 Revision of lead [electrode]
     - 37.76 Replacement of transvenous atrial and/or ventricular lead(s) [electrode]
     - 37.77 Removal of lead(s) [electrode] without replacement
     - 37.78 Insertion of temporary transvenous pacemaker system

2. **Rheumatic**

   - 390-392 Acute Rheumatic Fever
   - 393-398 Chronic Rheumatic Heart Disease

3. **Renal**
• 580 Acute glomerulonephritis
• 581 Nephrotic syndrome
• 582 Chronic glomerulonephritis
• 583 Nephritis and nephropathy not specified as acute or chronic
• 584 Acute kidney failure
• 585 Chronic kidney disease (ckd)
• 586 Renal failure, unspecified
• 4039.1  HTN CKD NOS V-ESRD
• 572.4 Hepatorenal syndrome

4. Neoplastic

• 140-149 Malignant Neoplasm Of Lip, Oral Cavity, And Pharynx
• 150-159 Malignant Neoplasm Of Digestive Organs And Peritoneum
• 160-165 Malignant Neoplasm Of Respiratory And Intrathoracic Organs
• 170-176 Malignant Neoplasm Of Bone, Connective Tissue, Skin, And Breast
• 179-189 Malignant Neoplasm Of Genitourinary Organs
• 190-199 Malignant Neoplasm Of Other And Unspecified Sites
• 200-209 Malignant Neoplasm Of Lymphatic And Hematopoietic Tissue
• 230-234 Carcinoma In Situ
• 235-238 Neoplasms Of Uncertain Behavior
• 239-239 Neoplasms Of Unspecified Nature
• 212 Benign neoplasm of respiratory and intrathoracic organs
• 228.1 Lymphangioma, any site
• 211.8 Benign neoplasm of retroperitoneum and peritoneum
• 229.0 Benign neoplasm of lymph nodes

5. Organ Transplant
- V42.0 Kidney replaced by transplant
- V42.1 Heart replaced by transplant
- V42.4 Bone replaced by transplant
- V42.6 Lung replaced by transplant
- V42.7 Liver replaced by transplant
- V42.81 Bone marrow replaced by transplant
- V42.82 Peripheral stem cells replaced by transplant
- V42.83 Pancreas replaced by transplant
- V42.84 Organ or tissue replaced by transplant, intestines
- V42.89 Other specified organ or tissue replaced by transplant
- V42.9 Unspecified organ or tissue replaced by transplant
- 996.8 Complications of transplanted organ
- 279.50 Graft-versus-host disease, unspecified
- 279.51 Acute graft-versus-host disease
- 279.52 Chronic graft-versus-host disease
- 279.53 Acute on chronic graft-versus-host disease

**Procedure code for:**

- 41.0 Bone marrow or hematopoietic stem cell transplant
- 37.51 Heart transplantation
- 33.6 Combined heart-lung transplantation
- 33.5 - Bilat lung transplant
- 50.5 Liver transplant
- 52.8 Transplant of pancreas
- 46.97 Transplant of intestine
- 55.6 Transplant of kidney
6. **Rheumatologic**

- 710 Diffuse diseases of connective tissue
- 713 Arthropathy associated with other disorders classified elsewhere
- 714 Rheumatoid arthritis and other inflammatory polyarthropathies
- 710.0 Systemic lupus erythematosus
- 710.1 Systemic sclerosis
- 710.2 Sicca syndrome
- 710.3 Dermatomyositis
- 710.4 Polymyositis
- 710.8 Other specified diffuse diseases of connective tissue
- 710.9 Unspecified diffuse connective tissue disease
- 277.31 Familial Mediterranean fever
- 695.4 Lupus erythematous
- 711.2 Arthropathy in behcet's syndrome
- 556 Ulcerative enterocolitis
- 040.2 Whipple's disease
- 555 Regional enteritis
- 696.0 Psoriatic arthropathy

7. **Kawasaki Disease**

- 446.1 Acute febrile mucocutaneous lymph node syndrome [MCLS]

8. **Vasculitis**

- 446.0 Polyarteritis nodosa
- 446.20 Hypersensitivity angiitis, unspecified
• 446.21 Goodpasture's syndrome
• 446.29 Other specified hypersensitivity angiitis
• 446.3 Lethal midline granuloma
• 446.4 Wegener's granulomatosis
• 446.5 Giant cell arteritis
• 446.6 Thrombotic microangiopathy
• 446.7 Takayasu's disease
• 447.6 Arteritis, unspecified
• 136.1 Behcet's syndrome
• 135 Sarcoidosis
• 287.0 Allergic purpura
• 695.13 Stevens-Johnson syndrome

9. Endocrine

• 242 Thyrotoxicosis with or without goiter
• 243 Congenital hypothyroidism
• 244 Acquired hypothyroidism
• 245 Thyroiditis
• 255.41 Glucocorticoid deficiency
• 036.3 Waterhouse-Friderichsen syndrome
• 250.1 Diabetes with ketoacidosis

10. Immunologic

• 279.00 Hypogammaglobulinemia, unspecified
• 279.01 Selective IgA immunodeficiency
• 279.02 Selective IgM immunodeficiency
• 279.03 Other selective immunoglobulin deficiencies
• 279.04 Congenital hypogammaglobulinemia
• 279.05 Immunodeficiency with increased IgM
• 279.06 Common variable immunodeficiency
• 279.09 Other deficiency of humoral immunity
• 279.10 Immunodeficiency with predominant T-cell defect, unspecified
• 279.11 Di George’s syndrome
• 279.12 Wiskott-aldrich syndrome
• 279.13 Nezelof’s syndrome
• 279.19 Other deficiency of cell-mediated immunity
• 279.2 Combined immunity deficiency
• 279.3 Unspecified immunity deficiency
• 279.4 Autoimmune disease not elsewhere classified
• 279.41 Autoimmune lymphoproliferative syndrome
• 279.49 Autoimmune disease, not elsewhere classified
• 279.8 Other specified disorders involving the immune mechanism
• 279.9 Unspecified disorder of immune mechanism
• 288.4 Hemophagocytic syndromes

11. Metabolic/Other

• 274 Gout
• 277.39 Other amyloidosis
• 307.1 Anorexia nervosa
• 265.0 Beriberi
• 260 Kwashiorkor
• 261 Nutritional marasmus
• 262 Other severe protein-calorie malnutrition
• 263 Other and unspecified protein-calorie malnutrition
• 995.0 Other anaphylactic reaction
• 995.6 Anaphylactic reaction due to unspec food
• 999.4 Anaphylactic reaction to serum
• 999.5 Other serum reaction not elsewhere classified
• 571 Chronic liver disease and cirrhosis
• 751.61 Biliary atresia

12. Trauma chest/mediastinum

• 860 Traumatic pneumothorax and hemothorax
• 861 Injury to heart and lung
• 862 Injury to other and unspecified intrathoracic organs
• 863 Injury to gastrointestinal tract
• 423.0 Hemopericardium
• 864 Injury to liver
• 865 Injury to spleen
• 994.0 Effects of lightning
• 994.8 Electrocution and nonfatal effects of electric current

13. Perinatal

• 773 Hemolytic disease of fetus or newborn due to isoimmunization
• 7780 Hydrops fetalis isoimm
• 74783 Persistent fetal circulation
• 776.2 Disseminated intravascular coagulation in newborn
• 768.73 Severe hypoxic-ischemic encephalopathy
• 768.72 Moderate hypoxic-ischemic encephalopathy
• 768.5 Severe birth asphyxia
• 772.14 Intraventricular hemorrhage, grade IV
• 772.13 Intraventricular hemorrhage, grade III
• 765.00 Extreme immaturity, unspecified [weight]
• 765.01 Extreme immaturity, less than 500 grams
• 765.02 Extreme immaturity, 500-749 grams
• 765.03 Extreme immaturity, 750-999 grams
• 765.04 Extreme immaturity, 1,000-1,249 grams
• 765.05 Extreme immaturity, 1,250-1,499 grams
• 765.06 Extreme immaturity, 1,500-1,749 grams
• 765.07 Extreme immaturity, 1,750-1,999 grams
• 765.08 Extreme immaturity, 2,000-2,499 grams
• 765.1 Disorders relating to other preterm infants
• 765.10 Other preterm infants, unspecified [weight]
• 765.11 Other preterm infants, less than 500 grams
• 765.12 Other preterm infants, 500-749 grams
• 765.13 Other preterm infants, 750-999 grams
• 765.14 Other preterm infants, 1,000-1,249 grams
• 765.15 Other preterm infants, 1,250-1,499 grams
• 765.16 Other preterm infants, 1,500-1,749 grams
• 765.17 Other preterm infants, 1,750-1,999 grams
• 765.18 Other preterm infants, 2,000-2,499 grams
• 765.21 Less than 24 completed weeks of gestation
• 765.22 24 completed weeks of gestation
• 765.23 25-26 completed weeks of gestation
• 765.24 27-28 completed weeks of gestation
• 765.25 29-30 completed weeks of gestation
• 765.26 31-32 completed weeks of gestation
• 765.27 33-34 completed weeks of gestation
• 765.28 35-36 completed weeks of gestation

14. Hematologic

• 283.0 Autoimmune hemolytic anemia
• 283.1 Non-autoimmune hemolytic anemias
• 283.10 Non-autoimmune hemolytic anemia, unspecified
• 283.11 Hemolytic-uremic syndrome
• 283.19 Other non-autoimmune hemolytic anemias
• 283.2 Hemoglobinuria due to hemolysis from external causes
• 283.9 Acquired hemolytic anemia, unspecified
• 286 Coagulation defects
• 282.6 Sickle-cell disease

15. Chronic Pericarditis

• 423.2 Calcareous, constrictive pericarditis, Concato’s disease, Pick’s disease of heart (and liver)
• 423.1 Adhesive, adherent, fibrous, obliterating, plastic pericarditis

16. Endocarditis

• 421.0 Acute and subacute bacterial endocarditis
• 421.1 Acute and subacute infective endocarditis in diseases classified elsewhere
• 421.9 Acute endocarditis, unspecified
• 112.81 Candidal endocarditis
• 424.90 Endocarditis, valve unspecified, unspecified cause
• 424.91 Endocarditis in diseases classified elsewhere
• 424.99 Other endocarditis, valve unspecified

17. Myocarditis

• 422.0 in diseases classified elsewhere
• 422.90 Acute myocarditis, unspecified
• 422.91 Idiopathic myocarditis
• 422.92 Septic myocarditis
• 422.93 Toxic myocarditis
• 422.99 Other acute myocarditis
• 429.0 Myocarditis, unspecified

18. Other non-viral Infections

• 115 Histoplasmosis
• 114 Coccidioidomycosis
• 116 Blastomycotic infection
• 112.4 Candidiasis of lung
• 112.5 Disseminated candidiasis
• 112.83 Candidal meningitis
• 117.5 Cryptococcosis
• 117.3 Aspergillosis
• 130 Toxoplasmosis
• 122 Echinococcosis
• 120 Schistosomiasis (bilharziasis)
• 100 Leptospirosis
• 125 Filarial infection and dracontiasis
• 124 Trichinosis
• 010-018 Tuberculosis
• 020-027 Zoonotic Bacterial Diseases
• 030-041 Other Bacterial Diseases
• 090-099 Syphilis And Other Venereal Diseases
• 100-104 Other Spirochetal Diseases
• 599.0 Urinary tract infection, site not specified
• 604 Orchitis and epididymitis
• 681 Cellulitis and abscess of finger and toe
• 682 Other cellulitis and abscess
• 683 Acute lymphadenitis
• 080-088 Rickettsioses And Other Arthropod-Borne Diseases
• 771.81 Septicemia of newborn
• 995.91 Sepsis
• 031 Diseases due to other mycobacteria
• 478.24 Retropharyngeal abscess
• 478.22 Parapharyngeal abscess
• 475 Peritonsillar abscess
• 996.6 Infection and inflammatory reaction due to internal prosthetic device implant and graft
• 999.3 Other infection due to medical care not elsewhere classified
• 569.5 Abscess of intestine
• 569.61 Infection of colostomy or enterostomy
• 566 Abscess of anal and rectal regions
• 008.45 Intestinal infection due to Clostridium difficile
• 008.5 Bacterial enteritis, unspecified
• 008.0 Intestinal infection due to escherichia coli [e. coli]
• 008.42 Intestinal infection due to pseudomonas
• 008.43 Intestinal infection due to campylobacter
• 008.44 Intestinal infection due to yersinia enterocolitica
• 008.46 Intestinal infection due to other anaerobes
• 008.47 Intestinal infection due to other gram-negative bacteria
• 002 Typhoid and paratyphoid fevers
• 006 Amebiasis
• 003 Other salmonella infections
• 001 Cholera

19. Contiguous non-viral Infection

• 006.3 Amebic liver abscess
• 530.86 Infection of esophagostomy
• 006.4 Amebic lung abscess
• 483 Pneumonia due to other specified organism
• 484.8 Pneumonia in other infectious diseases classified elsewhere
• 510 Empyema
• 513 Abscess of lung and mediastinum
• 484.3 Pneumonia in whooping cough
• 484.5 Pneumonia in anthrax
• 486 Pneumonia, organism unspecified
• 875 Open wound of chest (wall)
• 481 Pneumococcal pneumonia [Streptococcus pneumoniae pneumonia]
• 482 Other bacterial pneumonia
- 484.6 Pneumonia in aspergillosis
- 484.7 Pneumonia in other systemic mycoses
- 420.99 Bacterial, purulent, septic pericarditis, pyopericardium, pneumopyopericardium
- 115.03 Infection by Histoplasma capsulatum, pericarditis
- 115.13 Infection by Histoplasma duboisi, pericarditis
- 115.93 Histoplasmosis, unspecified pericarditis

20. Other underlying cardiac

- 410-414 Ischemic Heart Disease
- 415-417 Diseases of Pulmonary Circulation
- 430-438 Cerebrovascular Disease
- 451-459 Diseases Of Veins And Lymphatics, And Other Diseases Of Circulatory System
- 425 Cardiomyopathy
- 426 Conduction disorders
- 427 Cardiac dysrhythmias
- 428 Heart failure
- 429 Ill-defined descriptions and complications of heart disease
- 997.1 Cardiac complications, not elsewhere classified
- 441 Aortic aneurysm and dissection
- 996.61 Infection and inflammatory reaction due to cardiac device, implant, and graft

21. Congenital cardiac

- 746 Other congenital anomalies of heart
- 745 Bulbus cordis anomalies and anomalies of cardiac septal closure
- 747.0 Patent ductus arteriosus
- 747.1 Coarctation of aorta
* 747.2 Other congenital anomalies of aorta
* 747.3 Congenital anomalies of pulmonary artery
* 747.4 Congenital anomalies of great veins

**22. Contiguous Inflammation**

* 511 Pleurisy
* 512 Pneumothorax and air leak
* 507 Pneumonitis due to solids and liquids
* 519.2 Mediastinitis
* 567 Peritonitis and retroperitoneal infections
* 530.4 Perforation of esophagus
* 530.1 Esophagitis
* 531.1 Acute gastric ulcer with perforation
* 531.2 Acute gastric ulcer with hemorrhage and perforation
* 531.5 Chronic or unspecified gastric ulcer with perforation
* 531.6 Chronic or unspecified gastric ulcer with hemorrhage and perforation
* 532.1 Acute duodenal ulcer with perforation
* 532.2 Acute duodenal ulcer with hemorrhage and perforation
* 532.5 Chronic or unspecified duodenal ulcer with perforation
* 532.6 Chronic or unspecified duodenal ulcer with hemorrhage and perforation
* 533.1 Acute peptic ulcer of unspecified site with perforation
* 533.2 Acute peptic ulcer of unspecified site with hemorrhage and perforation
* 533.5 Chronic or unspecified peptic ulcer of unspecified site with perforation
* 533.6 Chronic or unspecified peptic ulcer of unspecified site with hemorrhage and perforation
* 534.1 Acute gastrojejunal ulcer with perforation
* 534.2 Acute gastrojejunal ulcer with hemorrhage and perforation
• 534.5 Chronic or unspecified gastrojejunal ulcer with perforation
• 534.6 Chronic or unspecified gastrojejunal ulcer with hemorrhage and perforation
• 569.83 Perforation of intestine
• 577.0 Acute pancreatitis
• 530.85 Barrett's esophagus
• 530.0 Achalasia and cardiospasm
• 540 Acute appendicitis
<table>
<thead>
<tr>
<th>Comprehensive Category</th>
<th>Primary Etiology Category</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rheumatologic</td>
<td>Rheumatologic</td>
</tr>
<tr>
<td></td>
<td>Vasculitis</td>
</tr>
<tr>
<td></td>
<td>Kawasaki disease</td>
</tr>
<tr>
<td>Metabolic</td>
<td>Endocrine</td>
</tr>
<tr>
<td></td>
<td>Metabolic/Other</td>
</tr>
<tr>
<td>Heme-Immune</td>
<td>Hematologic</td>
</tr>
<tr>
<td></td>
<td>Immunologic</td>
</tr>
<tr>
<td>Contiguous</td>
<td>Trauma</td>
</tr>
<tr>
<td></td>
<td>Gastrointestinal</td>
</tr>
<tr>
<td></td>
<td>chest/mediastinum</td>
</tr>
<tr>
<td>Cardiac</td>
<td>Cardiac</td>
</tr>
<tr>
<td></td>
<td>Congenital</td>
</tr>
<tr>
<td></td>
<td>Other underlying cardiac</td>
</tr>
<tr>
<td></td>
<td>Rheumatic</td>
</tr>
<tr>
<td></td>
<td>Other underlying cardiac</td>
</tr>
<tr>
<td></td>
<td>Non-viral non cardiac</td>
</tr>
<tr>
<td></td>
<td>Infections</td>
</tr>
<tr>
<td>Cardiac infection</td>
<td>Myocarditis</td>
</tr>
<tr>
<td></td>
<td>Endocarditis</td>
</tr>
<tr>
<td>Chronic pericarditis</td>
<td></td>
</tr>
<tr>
<td>Non-viral non cardiac</td>
<td></td>
</tr>
<tr>
<td>Infections</td>
<td></td>
</tr>
<tr>
<td>Organ Transplant</td>
<td></td>
</tr>
<tr>
<td>Neoplastic</td>
<td></td>
</tr>
<tr>
<td>Renal</td>
<td></td>
</tr>
<tr>
<td>Perinatal</td>
<td></td>
</tr>
</tbody>
</table>
Supplementary Table 4: Distribution of excluded patients with potential causes for pericarditis according to a hierarchical model*

<table>
<thead>
<tr>
<th>Category</th>
<th>N</th>
<th>(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Neoplastic</td>
<td>1416</td>
<td>(13.1)</td>
</tr>
<tr>
<td>2. Renal</td>
<td>1391</td>
<td>(12.9)</td>
</tr>
<tr>
<td>3. Rheumatologic (rheumatologic disorders, vasculitis, Kawasaki disease)</td>
<td>569</td>
<td>(5.3)</td>
</tr>
<tr>
<td>4. Organ Transplant</td>
<td>409</td>
<td>(3.8)</td>
</tr>
<tr>
<td>5. Cardiac Infection (myocarditis and endocarditis)</td>
<td>291</td>
<td>(2.7)</td>
</tr>
<tr>
<td>6. Cardiac (rheumatic, cardiac surgery and congenital heart disease)</td>
<td>5800</td>
<td>(53.7)</td>
</tr>
<tr>
<td>7. Contiguous (chest trauma, contiguous infection or inflammation)</td>
<td>662</td>
<td>(6.1)</td>
</tr>
<tr>
<td>8. Non-viral non-cardiac Infections</td>
<td>134</td>
<td>(1.2)</td>
</tr>
<tr>
<td>9. Heme-Immune</td>
<td>30</td>
<td>(0.3)</td>
</tr>
<tr>
<td>10. Metabolic (metabolic, endocrine)</td>
<td>40</td>
<td>(0.4)</td>
</tr>
<tr>
<td>11. Perinatal</td>
<td>52</td>
<td>(0.5)</td>
</tr>
<tr>
<td>12. Chronic pericarditis</td>
<td>8</td>
<td>(0.1)</td>
</tr>
</tbody>
</table>

* If a patient had ICD-9 codes for more than one category of possible etiology for pericarditis/effusion, the above hierarchical model was used to attribute an etiology. Category 1 has highest weight followed by the one below in sequential order.
Idiopathic Pericarditis and Pericardial Effusion in Children: Contemporary Epidemiology and Management

Divya Shakti, Rebecca Hehn, Kimberly Gauvreau, Robert P. Sundel and Jane W. Newburger

*J Am Heart Assoc.* 2014;3:e001483; originally published November 7, 2014;
doi: 10.1161/JAHA.114.001483

The *Journal of the American Heart Association* is published by the American Heart Association, 7272 Greenville Avenue,
Dallas, TX 75231
Online ISSN: 2047-9980

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://jaha.ahajournals.org/content/3/6/e001483

Data Supplement (unedited) at:
http://jaha.ahajournals.org/content/suppl/2014/12/01/jah3744.DC1
Supplementary Material

Supplementary Table 1: ICD-9 Diagnosis and Procedure codes along with their description, used for selection of discharges with pericarditis or pericardial effusion.

<table>
<thead>
<tr>
<th>ICD-9 Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Diagnosis Code</strong></td>
<td></td>
</tr>
<tr>
<td>423.9</td>
<td>Pericarditis (granular) (with decompensation) (with effusion), unspecified disease of pericardium</td>
</tr>
<tr>
<td>420.90</td>
<td>Acute (non-rheumatic) pericarditis, unspecified- hemorrhagic, infective NOS, neoplastic, sicca,</td>
</tr>
<tr>
<td>420.99</td>
<td>Bacterial, fibrinopurulent, fibrinous pericarditis- (acute) (subacute) (with serous or seropurulent effusion), pneumococcal, staphylococcal, streptococcal, suppurative, purulent, septic pericarditis, pyopericardium, pneumopyopericardium</td>
</tr>
<tr>
<td>420.91</td>
<td>Benign, idiopathic, non-specific, viral pericarditis</td>
</tr>
<tr>
<td>423.1</td>
<td>Adhesive, adherent, fibrous (external) (internal), obliterans, obliterating, plastic pericarditis, Milk spots, Soldier’s patches.</td>
</tr>
<tr>
<td>420.0</td>
<td>Acute pericarditis in diseases classified elsewhere (coded separately)</td>
</tr>
<tr>
<td>423.2</td>
<td>Calcareous, constrictive pericarditis, Concato’s disease, Pick’s disease of heart (and liver)</td>
</tr>
<tr>
<td>423.3</td>
<td>Cardiac tamponade</td>
</tr>
<tr>
<td>423.8</td>
<td>Chronic (non-rheumatic) neoplastic pericarditis, other unspecified diseases of pericardium</td>
</tr>
<tr>
<td>074.21</td>
<td>Coxsackie pericarditis</td>
</tr>
<tr>
<td>391.0</td>
<td>Acute rheumatic pericarditis</td>
</tr>
<tr>
<td>Code</td>
<td>Description</td>
</tr>
<tr>
<td>--------</td>
<td>-----------------------------------------------------------------------------</td>
</tr>
<tr>
<td>393</td>
<td>Inactive or quiescent (Chronic) Rheumatic –(pericarditis, mediastinopericarditis, myopericarditis)</td>
</tr>
<tr>
<td>115.03</td>
<td>Infection by Histoplasma capsulatum, pericarditis</td>
</tr>
<tr>
<td>115.13</td>
<td>Infection by Histoplasma duboisii, pericarditis</td>
</tr>
<tr>
<td>115.13</td>
<td>Infection by Histoplasma duboisii, pericarditis</td>
</tr>
<tr>
<td>115.93</td>
<td>Histoplasmosis, unspecified pericarditis</td>
</tr>
<tr>
<td>098.83</td>
<td>Gonococcal pericarditis</td>
</tr>
<tr>
<td>423.0</td>
<td>Hemopericardium</td>
</tr>
<tr>
<td>036.41</td>
<td>Meningococcal pericarditis</td>
</tr>
<tr>
<td>093.81</td>
<td>Syphilitic pericarditis</td>
</tr>
<tr>
<td>420</td>
<td>Acute pericarditis (mediastinopericarditis, myopericarditis, pericardial effusion, pleuroperticarditis, pneumopericarditis)</td>
</tr>
<tr>
<td>420.9</td>
<td>Other and unspecified acute pericarditis</td>
</tr>
<tr>
<td>411.0</td>
<td>Dressler’s (post myocardial infarction syndrome)</td>
</tr>
</tbody>
</table>

**Procedure Codes**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>37.0</td>
<td>Pericardiocentesis</td>
</tr>
<tr>
<td>37.12</td>
<td>Pericardiotomy</td>
</tr>
<tr>
<td>37.31</td>
<td>Pericardiectomy</td>
</tr>
</tbody>
</table>
Supplementary Table 2: ICD-9 Codes categorized for possible etiologies for pericarditis

1. **Cardiac Surgery/Procedure**

   - RACHS-1 entry criteria codes

   **Procedure code:**
   - 39.6 Extracorporeal circulation and procedures auxiliary to heart surgery

   **Procedure codes for pacemaker placement:**
   - 37.7 Insertion, revision, replacement, and removal of leads; insertion of temporary pacemaker system; or revision of cardiac device pocket
   - 37.70 Initial insertion of lead [electrode], not otherwise specified
   - 37.71 Initial insertion of transvenous lead [electrode] into ventricle
   - 37.72 Initial insertion of transvenous leads [electrodes] into atrium and ventricle
   - 37.73 Initial insertion of transvenous lead [electrode] into atrium
   - 37.75 Revision of lead [electrode]
   - 37.76 Replacement of transvenous atrial and/or ventricular lead(s) [electrode]
   - 37.77 Removal of lead(s) [electrode] without replacement
   - 37.78 Insertion of temporary transvenous pacemaker system

2. **Rheumatic**

   - 390-392 Acute Rheumatic Fever
   - 393-398 Chronic Rheumatic Heart Disease

3. **Renal**
• 580 Acute glomerulonephritis
• 581 Nephrotic syndrome
• 582 Chronic glomerulonephritis
• 583 Nephritis and nephropathy not specified as acute or chronic
• 584 Acute kidney failure
• 585 Chronic kidney disease (ckd)
• 586 Renal failure, unspecified
• 4039.1 HTN CKD NOS V-ESRD
• 572.4 Hepatorenal syndrome

4. Neoplastic
• 140-149 Malignant Neoplasm Of Lip, Oral Cavity, And Pharynx
• 150-159 Malignant Neoplasm Of Digestive Organs And Peritoneum
• 160-165 Malignant Neoplasm Of Respiratory And Intrathoracic Organs
• 170-176 Malignant Neoplasm Of Bone, Connective Tissue, Skin, And Breast
• 179-189 Malignant Neoplasm Of Genitourinary Organs
• 190-199 Malignant Neoplasm Of Other And Unspecified Sites
• 200-209 Malignant Neoplasm Of Lymphatic And Hematopoietic Tissue
• 230-234 Carcinoma In Situ
• 235-238 Neoplasms Of Uncertain Behavior
• 239-239 Neoplasms Of Unspecified Nature
• 212 Benign neoplasm of respiratory and intrathoracic organs
• 228.1 Lymphangioma, any site
• 211.8 Benign neoplasm of retroperitoneum and peritoneum
• 229.0 Benign neoplasm of lymph nodes

5. Organ Transplant
• V42.0 Kidney replaced by transplant
• V42.1 Heart replaced by transplant
• V42.4 Bone replaced by transplant
• V42.6 Lung replaced by transplant
• V42.7 Liver replaced by transplant
• V42.81 Bone marrow replaced by transplant
• V42.82 Peripheral stem cells replaced by transplant
• V42.83 Pancreas replaced by transplant
• V42.84 Organ or tissue replaced by transplant, intestines
• V42.89 Other specified organ or tissue replaced by transplant
• V42.9 Unspecified organ or tissue replaced by transplant
• 996.8 Complications of transplanted organ
• 279.50 Graft-versus-host disease, unspecified
• 279.51 Acute graft-versus-host disease
• 279.52 Chronic graft-versus-host disease
• 279.53 Acute on chronic graft-versus-host disease

Procedure code for:

• 41.0 Bone marrow or hematopoietic stem cell transplant
• 37.51 Heart transplantation
• 33.6 Combined heart-lung transplantation
• 33.5 - Bilat lung transplant
• 50.5 Liver transplant
• 52.8 Transplant of pancreas
• 46.97 Transplant of intestine
• 55.6 Transplant of kidney
6. **Rheumatologic**

- 710 Diffuse diseases of connective tissue
- 713 Arthropathy associated with other disorders classified elsewhere
- 714 Rheumatoid arthritis and other inflammatory polyarthropathies
- 710.0 Systemic lupus erythematosus
- 710.1 Systemic sclerosis
- 710.2 Sicca syndrome
- 710.3 Dermatomyositis
- 710.4 Polymyositis
- 710.8 Other specified diffuse diseases of connective tissue
- 710.9 Unspecified diffuse connective tissue disease
- 277.31 Familial Mediterranean fever
- 695.4 Lupus erythematosus
- 711.2 Arthropathy in behcet's syndrome
- 556 Ulcerative enterocolitis
- 040.2 Whipple's disease
- 555 Regional enteritis
- 696.0 Psoriatic arthropathy

7. **Kawasaki Disease**

- 446.1 Acute febrile mucocutaneous lymph node syndrome [MCLS]

8. **Vasculitis**

- 446.0 Polyarteritis nodosa
- 446.20 Hypersensitivity angiitis, unspecified
• 446.21 Goodpasture's syndrome
• 446.29 Other specified hypersensitivity angiitis
• 446.3 Lethal midline granuloma
• 446.4 Wegener’s granulomatosis
• 446.5 Giant cell arteritis
• 446.6 Thrombotic microangiopathy
• 446.7 Takayasu’s disease
• 447.6 Arteritis, unspecified
• 136.1 Behcet's syndrome
• 135 Sarcoidosis
• 287.0 Allergic purpura
• 695.13 Stevens-Johnson syndrome

9. Endocrine

• 242 Thyrotoxicosis with or without goiter
• 243 Congenital hypothyroidism
• 244 Acquired hypothyroidism
• 245 Thyroiditis
• 255.41 Glucocorticoid deficiency
• 036.3 Waterhouse-Friderichsen syndrome
• 250.1 Diabetes with ketoacidosis

10. Immunologic

• 279.00 Hypogammaglobulinemia, unspecified
• 279.01 Selective IgA immunodeficiency
• 279.02 Selective IgM immunodeficiency
- 279.03 Other selective immunoglobulin deficiencies
- 279.04 Congenital hypogammaglobulinemia
- 279.05 Immunodeficiency with increased IgM
- 279.06 Common variable immunodeficiency
- 279.09 Other deficiency of humoral immunity
- 279.10 Immunodeficiency with predominant T-cell defect, unspecified
- 279.11 Di George’s syndrome
- 279.12 Wiskott-aldrich syndrome
- 279.13 Nezelof’s syndrome
- 279.19 Other deficiency of cell-mediated immunity
- 279.2 Combined immunity deficiency
- 279.3 Unspecified immunity deficiency
- 279.4 Autoimmune disease not elsewhere classified
- 279.41 Autoimmune lymphoproliferative syndrome
- 279.49 Autoimmune disease, not elsewhere classified
- 279.8 Other specified disorders involving the immune mechanism
- 279.9 Unspecified disorder of immune mechanism
- 288.4 Hemophagocytic syndromes

11. Metabolic/Other

- 274 Gout
- 277.39 Other amyloidosis
- 307.1 Anorexia nervosa
- 265.0 Beriberi
- 260 Kwashiorkor
- 261 Nutritional marasmus
• 262 Other severe protein-calorie malnutrition
• 263 Other and unspecified protein-calorie malnutrition
• 995.0 Other anaphylactic reaction
• 995.6 Anaphylactic reaction due to unspec food
• 999.4 Anaphylactic reaction to serum
• 999.5 Other serum reaction not elsewhere classified
• 571 Chronic liver disease and cirrhosis
• 751.61 Biliary atresia

12. Trauma chest/mediastinum

• 860 Traumatic pneumothorax and hemothorax
• 861 Injury to heart and lung
• 862 Injury to other and unspecified intrathoracic organs
• 863 Injury to gastrointestinal tract
• 423.0 Hemopericardium
• 864 Injury to liver
• 865 Injury to spleen
• 994.0 Effects of lightning
• 994.8 Electrocution and nonfatal effects of electric current

13. Perinatal

• 773 Hemolytic disease of fetus or newborn due to isoimmunization
• 7780 Hydrops fetalis isoimm
• 74783 Persistent fetal circulation
• 776.2 Disseminated intravascular coagulation in newborn
• 768.73 Severe hypoxic-ischemic encephalopathy
- 768.72 Moderate hypoxic-ischemic encephalopathy
- 768.5 Severe birth asphyxia
- 772.14 Intraventricular hemorrhage, grade IV
- 772.13 Intraventricular hemorrhage, grade III
- 765.00 Extreme immaturity, unspecified [weight]
- 765.01 Extreme immaturity, less than 500 grams
- 765.02 Extreme immaturity, 500-749 grams
- 765.03 Extreme immaturity, 750-999 grams
- 765.04 Extreme immaturity, 1,000-1,249 grams
- 765.05 Extreme immaturity, 1,250-1,499 grams
- 765.06 Extreme immaturity, 1,500-1,749 grams
- 765.07 Extreme immaturity, 1,750-1,999 grams
- 765.08 Extreme immaturity, 2,000-2,499 grams
- 765.1 Disorders relating to other preterm infants
- 765.10 Other preterm infants, unspecified [weight]
- 765.11 Other preterm infants, less than 500 grams
- 765.12 Other preterm infants, 500-749 grams
- 765.13 Other preterm infants, 750-999 grams
- 765.14 Other preterm infants, 1,000-1,249 grams
- 765.15 Other preterm infants, 1,250-1,499 grams
- 765.16 Other preterm infants, 1,500-1,749 grams
- 765.17 Other preterm infants, 1,750-1,999 grams
- 765.18 Other preterm infants, 2,000-2,499 grams
- 765.21 Less than 24 completed weeks of gestation
- 765.22 24 completed weeks of gestation
- 765.23 25-26 completed weeks of gestation
• 765.24 27-28 completed weeks of gestation
• 765.25 29-30 completed weeks of gestation
• 765.26 31-32 completed weeks of gestation
• 765.27 33-34 completed weeks of gestation
• 765.28 35-36 completed weeks of gestation

14. Hematologic

• 283.0 Autoimmune hemolytic anemia
• 283.1 Non-autoimmune hemolytic anemias
• 283.10 Non-autoimmune hemolytic anemia, unspecified
• 283.11 Hemolytic-uremic syndrome
• 283.19 Other non-autoimmune hemolytic anemias
• 283.2 Hemoglobinuria due to hemolysis from external causes
• 283.9 Acquired hemolytic anemia, unspecified
• 286 Coagulation defects
• 282.6 Sickle-cell disease

15. Chronic Pericarditis

• 423.2 Calcareous, constrictive pericarditis, Concato’s disease, Pick’s disease of heart (and liver)
• 423.1 Adhesive, adherent, fibrous, obliterating, plastic pericarditis

16. Endocarditis

• 421.0 Acute and subacute bacterial endocarditis
• 421.1 Acute and subacute infective endocarditis in diseases classified elsewhere
• 421.9 Acute endocarditis, unspecified
• 112.81 Candidal endocarditis
• 424.90 Endocarditis, valve unspecified, unspecified cause
• 424.91 Endocarditis in diseases classified elsewhere
• 424.99 Other endocarditis, valve unspecified

17. **Myocarditis**

• 422.0 in diseases classified elsewhere
• 422.90 Acute myocarditis, unspecified
• 422.91 Idiopathic myocarditis
• 422.92 Septic myocarditis
• 422.93 Toxic myocarditis
• 422.99 Other acute myocarditis
• 429.0 Myocarditis, unspecified

18. **Other non-viral Infections**

• 115 Histoplasmosis
• 114 Coccidioidomycosis
• 116 Blastomycotic infection
• 112.4 Candidiasis of lung
• 112.5 Disseminated candidiasis
• 112.83 Candidal meningitis
• 117.5 Cryptococcosis
• 117.3 Aspergillosis
• 130 Toxoplasmosis
• 122 Echinococcosis
• 120 Schistosomiasis (bilharziasis)
• 100 Leptospirosis
• 125 Filarial infection and dracunculiasis
• 124 Trichinosis
• 010-018 Tuberculosis
• 020-027 Zoonotic Bacterial Diseases
• 030-041 Other Bacterial Diseases
• 090-099 Syphilis And Other Venereal Diseases
• 100-104 Other Spirochetal Diseases
• 599.0 Urinary tract infection, site not specified
• 604 Orchitis and epididymitis
• 681 Cellulitis and abscess of finger and toe
• 682 Other cellulitis and abscess
• 683 Acute lymphadenitis
• 080-088 Rickettsioses And Other Arthropod-Borne Diseases
• 771.81 Septicemia of newborn
• 995.91 Sepsis
• 031 Diseases due to other mycobacteria
• 478.24 Retropharyngeal abscess
• 478.22 Parapharyngeal abscess
• 475 Peritonsillar abscess
• 996.6 Infection and inflammatory reaction due to internal prosthetic device implant and graft
• 999.3 Other infection due to medical care not elsewhere classified
• 569.5 Abscess of intestine
• 569.61 Infection of colostomy or enterostomy
• 566 Abscess of anal and rectal regions
• 008.45 Intestinal infection due to Clostridium difficile
• 008.5 Bacterial enteritis, unspecified
• 008.0 Intestinal infection due to escherichia coli [e. coli]
• 008.42 Intestinal infection due to pseudomonas
• 008.43 Intestinal infection due to campylobacter
• 008.44 Intestinal infection due to yersinia enterocolitica
• 008.46 Intestinal infection due to other anaerobes
• 008.47 Intestinal infection due to other gram-negative bacteria
• 002 Typhoid and paratyphoid fevers
• 006 Amebiasis
• 003 Other salmonella infections
• 001 Cholera

19. Contiguous non-viral Infection

• 006.3 Amebic liver abscess
• 530.86 Infection of esophagostomy
• 006.4 Amebic lung abscess
• 483 Pneumonia due to other specified organism
• 484.8 Pneumonia in other infectious diseases classified elsewhere
• 510 Empyema
• 513 Abscess of lung and mediastinum
• 484.3 Pneumonia in whooping cough
• 484.5 Pneumonia in anthrax
• 486 Pneumonia, organism unspecified
• 875 Open wound of chest (wall)
• 481 Pneumococcal pneumonia [Streptococcus pneumoniae pneumonia]
• 482 Other bacterial pneumonia
• 484.6 Pneumonia in aspergillosis
• 484.7 Pneumonia in other systemic mycoses
• 420.99 Bacterial, purulent, septic pericarditis, pyopericardium, pneumopyopericardium
• 115.03 Infection by Histoplasma capsulatum, pericarditis
• 115.13 Infection by Histoplasma duboisii, pericarditis
• 115.93 Histoplasmosis, unspecified pericarditis

20. Other underlying cardiac

• 410-414 Ischemic Heart Disease
• 415-417 Diseases of Pulmonary Circulation
• 430-438 Cerebrovascular Disease
• 451-459 Diseases Of Veins And Lymphatics, And Other Diseases Of Circulatory System
• 425 Cardiomyopathy
• 426 Conduction disorders
• 427 Cardiac dysrhythmias
• 428 Heart failure
• 429 Ill-defined descriptions and complications of heart disease
• 997.1 Cardiac complications, not elsewhere classified
• 441 Aortic aneurysm and dissection
• 996.61 Infection and inflammatory reaction due to cardiac device, implant, and graft

21. Congenital cardiac

• 746 Other congenital anomalies of heart
• 745 Bulbus cordis anomalies and anomalies of cardiac septal closure
• 747.0 Patent ductus arteriosus
• 747.1 Coarctation of aorta
• 747.2 Other congenital anomalies of aorta
• 747.3 Congenital anomalies of pulmonary artery
• 747.4 Congenital anomalies of great veins

22. Contiguous Inflammation

• 511 Pleurisy
• 512 Pneumothorax and air leak
• 507 Pneumonitis due to solids and liquids
• 519.2 Mediastinitis
• 567 Peritonitis and retroperitoneal infections
• 530.4 Perforation of esophagus
• 530.1 Esophagitis
• 531.1 Acute gastric ulcer with perforation
• 531.2 Acute gastric ulcer with hemorrhage and perforation
• 531.5 Chronic or unspecified gastric ulcer with perforation
• 531.6 Chronic or unspecified gastric ulcer with hemorrhage and perforation
• 532.1 Acute duodenal ulcer with perforation
• 532.2 Acute duodenal ulcer with hemorrhage and perforation
• 532.5 Chronic or unspecified duodenal ulcer with perforation
• 532.6 Chronic or unspecified duodenal ulcer with hemorrhage and perforation
• 533.1 Acute peptic ulcer of unspecified site with perforation
• 533.2 Acute peptic ulcer of unspecified site with hemorrhage and perforation
• 533.5 Chronic or unspecified peptic ulcer of unspecified site with perforation
• 533.6 Chronic or unspecified peptic ulcer of unspecified site with hemorrhage and perforation
• 534.1 Acute gastrojejunal ulcer with perforation
• 534.2 Acute gastrojejunal ulcer with hemorrhage and perforation
• 534.5 Chronic or unspecified gastrojejunal ulcer with perforation
• 534.6 Chronic or unspecified gastrojejunal ulcer with hemorrhage and perforation
• 569.83 Perforation of intestine
• 577.0 Acute pancreatitis
• 530.85 Barrett's esophagus
• 530.0 Achalasia and cardiospasm
• 540 Acute appendicitis
**Supplementary Table 3: Categorization of possible etiologies.**

<table>
<thead>
<tr>
<th>Comprehensive Category</th>
<th>Primary Etiology Category</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rheumatologic</td>
<td>Rheumatologic</td>
</tr>
<tr>
<td></td>
<td>Vasculitis</td>
</tr>
<tr>
<td></td>
<td>Kawasaki disease</td>
</tr>
<tr>
<td>Metabolic</td>
<td>Endocrine</td>
</tr>
<tr>
<td></td>
<td>Metabolic/Other</td>
</tr>
<tr>
<td>Heme-Immune</td>
<td>Hematologic</td>
</tr>
<tr>
<td></td>
<td>Immunologic</td>
</tr>
<tr>
<td>Contiguous</td>
<td>Trauma</td>
</tr>
<tr>
<td>chest/mediastinum</td>
<td>Gastrointestinal</td>
</tr>
<tr>
<td>Cardiac</td>
<td>Cardiac surgery\procedure</td>
</tr>
<tr>
<td></td>
<td>cardiac</td>
</tr>
<tr>
<td></td>
<td>Other underlying cardiac</td>
</tr>
<tr>
<td>Cardiac infection</td>
<td>Myocarditis</td>
</tr>
<tr>
<td></td>
<td>Endocarditis</td>
</tr>
<tr>
<td>Chronic pericarditis</td>
<td></td>
</tr>
<tr>
<td>Non-viral non cardiac Infections</td>
<td>Infections</td>
</tr>
<tr>
<td>Organ Transplant</td>
<td></td>
</tr>
<tr>
<td>Neoplastic</td>
<td></td>
</tr>
<tr>
<td>Renal</td>
<td></td>
</tr>
<tr>
<td>Perinatal</td>
<td></td>
</tr>
</tbody>
</table>
### Supplementary Table 4: Distribution of excluded patients with potential causes for pericarditis according to a hierarchical model*

<table>
<thead>
<tr>
<th>Category</th>
<th>N</th>
<th>(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Neoplastic</td>
<td>1416</td>
<td>13.1</td>
</tr>
<tr>
<td>2. Renal</td>
<td>1391</td>
<td>12.9</td>
</tr>
<tr>
<td>3. Rheumatologic (rheumatologic disorders, vasculitis, Kawasaki disease)</td>
<td>569</td>
<td>5.3</td>
</tr>
<tr>
<td>4. Organ Transplant</td>
<td>409</td>
<td>3.8</td>
</tr>
<tr>
<td>5. Cardiac Infection (myocarditis and endocarditis)</td>
<td>291</td>
<td>2.7</td>
</tr>
<tr>
<td>6. Cardiac (rheumatic, cardiac surgery and congenital heart disease)</td>
<td>5800</td>
<td>53.7</td>
</tr>
<tr>
<td>7. Contiguous (chest trauma, contiguous infection or inflammation)</td>
<td>662</td>
<td>6.1</td>
</tr>
<tr>
<td>8. Non-viral non-cardiac Infections</td>
<td>134</td>
<td>1.2</td>
</tr>
<tr>
<td>9. Heme-Immune</td>
<td>30</td>
<td>0.3</td>
</tr>
<tr>
<td>10. Metabolic (metabolic, endocrine)</td>
<td>40</td>
<td>0.4</td>
</tr>
<tr>
<td>11. Perinatal</td>
<td>52</td>
<td>0.5</td>
</tr>
<tr>
<td>12. Chronic pericarditis</td>
<td>8</td>
<td>0.1</td>
</tr>
</tbody>
</table>

* If a patient had ICD-9 codes for more than one category of possible etiology for pericarditis/effusion, the above hierarchical model was used to attribute an etiology. Category 1 has highest weight followed by the one below in sequential order.