



## Cardiovascular Complications are Common in Patients with Juvenile Dermatomyositis in a Cross-Sectional Analysis of the 2016 Kids Inpatient Database

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### Introduction

Juvenile dermatomyositis (JDM) is an uncommon inflammatory disease (incidence 2-4 per 1 million children) with frequent cardiovascular complications (CVCs). Incidence of CVCs in JDM varied widely from 2.9% to 37.5% in a meta-analysis of single-institution studies [1]. We aimed to assess for differences in presentation, treatment, and outcomes of JDM pediatric inpatients with CVCs.

The 2016 Kids Inpatient Database (KID), an all-payer in-patient pediatric database [2], was queried for JDM patients using International Classification of Diseases, Tenth Revision (ICD-10), Clinical Modification JDM code “M33.” CVCs were collected using codes “A52.0/E08–E16/E66/E78/100–I99/Q2/Z95.” Chi-squared tests compared frequencies

by CVC status. Multivariable logistic regression identified predictors of CVCs ( $P < 0.05$ ).

### Case Presentation

There were 836 pediatric JDM cases and 222 (26.6%) had CVCs (Table 1). Patients with CVCs were older (14.0 versus 11.4 years) and more often Black (29.1% versus 21.6%), Hispanic (35.5% versus 27.7%), in the lowest income quartile (36.1% versus 29.3%), and Medicaid-insured (52.3% versus 41.9%) (all  $P < 0.05$ ). Overall treatment nonadherence was more common for patients ages 14-20 (71.4% versus 44.2%), females (96.4% versus 78.1%), and Blacks (48.3% versus 22.7%) ( $P < 0.025$ ) (Supplementary Table 1).

**Table 1. Demographic data of dermatomyositis by cardiovascular complication status.**

|   |   | No Cardiovascular Complications | Cardiovascular Complications | Total        | P       |
|---|---|---------------------------------|------------------------------|--------------|---------|
|   |   | N = 613 (73.4%)                 | N = 222 (26.6%)              | N = 836      |         |
| Age   | Age, years (mean [SE])                          | 11.42 [0.21]                    | 14.04 [0.31]                 | 12.12 [0.18] | < 0.001 |
| Sex   | Male  | 22.2%                           | 19.3%                        | 21.4%        | 0.365   |
|   | Female  | 77.8%                           | 80.7%                        | 78.6%        |         |
| Race  | White   | 44.5%                           | 29.1%                        | 40.4%        | 0.002   |
|   | Black   | 21.6%                           | 29.1%                        | 23.6%        |         |
|   | Hispanic  | 27.7%                           | 35.5%                        | 29.8%        |         |
|   | Other   | 6.1%                            | 6.4%                         | 6.2%         |         |
| Median Income Quartile - Patient Zip Code       | 0 – 25%   | 29.3%                           | 36.1%                        | 31.1%        | 0.038   |
|   | 26 – 50%  | 25.6%                           | 25.1%                        | 25.5%        |         |
|   | 51 – 75%  | 24.6%                           | 26.5%                        | 25.1%        |         |
|   | 76 – 100%                                       | 20.5%                           | 12.3%                        | 18.3%        |         |
| Primary Payer Status                            | Medicare  | 0.2%                            | 1.4%                         | 0.5%         | 0.005   |
|   | Medicaid  | 41.9%                           | 52.3%                        | 44.7%        |         |
|   | Private Insurance                               | 49.4%                           | 39.6%                        | 46.8%        |         |
|   | Self-Pay  | 2.8%                            | 0.5%                         | 2.2%         |         |
|   | No Charge                                       | 0.2%                            | 0.0%                         | 0.1%         |         |
|   | Other   | 5.5%                            | 6.3%                         | 5.7%         |         |
| Hospital Region                                 | Northeast                                       | 15.2%                           | 8.1%                         | 13.3%        | < 0.001 |
|   | Midwest   | 22.5%                           | 15.8%                        | 20.7%        |         |
|   | South   | 40.6%                           | 40.1%                        | 40.5%        |         |
|   | West  | 21.7%                           | 36.0%                        | 25.5%        |         |
| Severity of Illness Subclass (Loss of Function) | Minor LOF                                       | 36.4%                           | 10.3%                        | 29.4%        | < 0.001 |
|   | Moderate LOF                                    | 43.6%                           | 43.5%                        | 43.5%        |         |
|   | Major LOF                                       | 16.0%                           | 34.5%                        | 20.9%        |         |
|   | Extreme LOF                                     | 4.1%                            | 11.7%                        | 6.1%         |         |
| Comorbidity                                     | Anemia  | 15.2%                           | 27.5%                        | 18.4%        | < 0.001 |
|   | Fluid & Electrolyte Disorder                    | 10.8%                           | 23.3%                        | 14.1%        | < 0.001 |
|   | Aphagia & Dysphagia                             | 9.5%                            | 6.8%                         | 8.7%         | 0.222   |
|   | Asthma  | 7.7%                            | 10.8%                        | 8.5%         | 0.154   |
|   | Heartbeat Abnormalities at Initial Presentation | 5.1%                            | 11.7%                        | 6.8%         | < 0.001 |
|   | Coagulation Defect                              | 4.2%                            | 12.6%                        | 6.5%         | < 0.001 |
|   | Gastro-esophageal Reflux Disease                | 3.4%                            | 11.3%                        | 5.5%         | < 0.001 |
|   | Liver Disease                                   | 1.6%                            | 6.8%                         | 2.9%         | < 0.001 |
|   | Perinatal Chronic Respiratory Disease           | 0.7%                            | 3.2%                         | 1.3%         | 0.005   |
|   | Treatment Nonadherence                          | 1.6%                            | 8.1%                         | 3.4%         | < 0.001 |

LOF = loss of function; SE = standard error.

JDM patients with vs. without CVCs had higher incidence of acute kidney injury (AKI) (5.8% versus 1.3%,  $P < 0.001$ ) (Table 2). On multivariable analysis, CVCs were associated with increasing age (OR 1.12, 95% CI 1.07–1.16) and heartbeat abnormalities at initial presentation (OR 2.67, 95% CI 1.37–5.17) ( $P < 0.005$ ) (Supplementary Table 2).

## Conclusions

We found that JDM inpatients with CVCs were most often Black or Hispanic, of lower income, and Medicaid-insured.

Similarly, in a national study of 16,097 pediatric inpatients, Blacks vs. Whites were 20% more likely to die within 30 days of surgery, which were attributed to lack of specialized care access, poor physician-parent communication, and systemic racism [3]. Therefore, social determinants likely influence the development of CVCs in JDM patients.

We found that JDM patients with CVCs had greater total charges, LOS, and incidence of AKI, but no difference in the number of procedures performed, suggesting worse inpatient disease courses for JDM patients with CVCs. In a 10-year

**Supplementary Table 1. Demographics of dermatomyositis by treatment adherence status.**

|      |             | Treatment Adherence | Treatment Nonadherence | Total   | P     |
|------|-------------|---------------------|------------------------|---------|-------|
|      |             | N = 808 (96.6%)     | N = 28 (3.4%)          | N = 836 |       |
| Age  | 0-6 years   | 17.9%               | 3.6%                   | 17.5%   | 0.012 |
|      | 7-13 years  | 37.9%               | 25.0%                  | 37.4%   |       |
|      | 14-20 years | 44.2%               | 71.4%                  | 45.1%   |       |
| Sex  | Male        | 21.9%               | 3.6%                   | 21.3%   | 0.020 |
|      | Female      | 78.1%               | 96.4%                  | 78.7%   |       |
| Race | White       | 41.1%               | 20.7%                  | 40.3%   | 0.006 |
|      | Black       | 22.7%               | 48.3%                  | 23.7%   |       |
|      | Hispanic    | 29.8%               | 31.0%                  | 29.8%   |       |
|      | Other       | 6.4%                | 0.0%                   | 6.2%    |       |

**Table 2. Management and outcomes of dermatomyositis by cardiovascular complication status.**

|                                      |                                  | No Cardiovascular Complications | Cardiovascular Complications | Total                | P       |
|--------------------------------------|----------------------------------|---------------------------------|------------------------------|----------------------|---------|
| Total Charges                        | Charges (\$) (mean [SE])         | 52,432.22 [3,579.96]            | 110,743.53 [15,989.20]       | 67,956.85 [5,076.36] | < 0.001 |
| Length of Stay                       | Number of Days (mean [SE])       | 4.51 [0.34]                     | 8.54 [1.07]                  | 5.58 [0.39]          | < 0.001 |
| Number of Procedures                 | Number of Procedures (mean [SE]) | 1.27 [0.08]                     | 1.58 [0.19]                  | 1.36 [0.08]          | 0.137   |
| Time Until 1 <sup>st</sup> Procedure | Number of Days (mean [SE])       | 2.00 [0.46]                     | 2.81 [0.42]                  | 2.21 [0.36]          | 0.320   |
| Sepsis                               | Complication (%)                 | 4.7%                            | 4.0%                         | 4.5%                 | 0.670   |
| Respiratory Failure                  | Complication (%)                 | 3.1%                            | 5.0%                         | 3.6%                 | 0.202   |
| Urinary Tract Infection              | Complication (%)                 | 2.4%                            | 4.5%                         | 3.0%                 | 0.123   |
| Acute Kidney Injury                  | Complication (%)                 | 1.3%                            | 5.8%                         | 2.5%                 | < 0.001 |
| Hypoxemia                            | Complication (%)                 | 0.5%                            | 2.7%                         | 1.1%                 | 0.006   |
| Mortality                            | Complication (%)                 | 0.2%                            | 0.0%                         | 0.1%                 | 0.547   |
| Transfusion                          | Procedure (%)                    | 20.2%                           | 17.1%                        | 19.4%                | 0.320   |
| Imaging                              | Procedure (%)                    | 10.1%                           | 7.7%                         | 9.5%                 | 0.284   |

SE = standard error.

**Supplementary Table 2. Binary logistic regression analysis of factors associated with cardiovascular complications in dermatomyositis.**

|   | Odds Ratio | 95% CI     | P <sup>a</sup> |
|---|------------|------------|----------------|
| Age   | 1.12       | 1.07-1.16  | < 0.001        |
| West vs. Northeast                              | 3.80       | 1.82-7.91  | < 0.001        |
| Fluid and Electrolyte Disorders                 | 1.93       | 1.14-3.28  | 0.015          |
| Heartbeat Abnormalities at Initial Presentation | 2.87       | 1.48-5.57  | 0.002          |
| Gastro-esophageal Reflux Disease                | 3.72       | 1.77-7.82  | < 0.001        |
| Perinatal Chronic Respiratory Disease           | 6.17       | 1.60-23.73 | 0.008          |
| Treatment Nonadherence                          | 5.31       | 1.98-14.22 | < 0.001        |

CI = confidence interval. <sup>a</sup>Multivariable analysis with age, race, income quartile, primary payer status, hospital region, anemia, fluid and electrolyte disorders, heartbeat abnormalities, coagulation defects, gastro-esophageal reflux disease, liver disease, perinatal chronic respiratory disease, and treatment nonadherence.

single institution registry study, 1992-2002, AKI incidence was 21.5% among 65 JDM patients, while overall incidence of AKI was 2.5%. Therefore, CVCs may be contributory to cost differences and LOS in JDM patients [4].

CVCs were associated with tachycardia, bradycardia, and palpitations at initial presentation. Continuous monitoring with wearables in multiple sclerosis patients accurately showed trend-based heart rate variability and general dysregulation [5]. Therefore, screening for heartbeat abnormalities might detect undiagnosed CVCs and preventing disease progression in JDM patients.

Older, female, and Black children had higher incidence of treatment nonadherence, which may explain the age and CVC association with multivariable analysis. Given the predominance of United States White dermatologists [6], physician/patient race discordance may be partially responsible for this nonadherence.

Limitations include retrospective design and lack of data regarding medications administered and procedures performed.

We conclude that for JDM patients with CVCs, there are significant disparities in income, race, and insurance status. Dermatologists treating pediatric JDM patients should screen for CVCs with appropriate cardiologist referral to improve outcomes for these patients.

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