Systemic Juvenile Idiopathic Arthritis Inactive Disease and Withdrawal of Medications: A Survey of the Childhood Arthritis and Rheumatology Research Alliance

Kabita Nanda[1], Susan Shenoi[1], Ashley Cooper[2], Bridget Edghill[3], Miriah Gillispie[4], Baruch Goldberg[5], Olha Halyabar[6], Daniel B. Horton[7], Thomas G. Mason[8], Tova Ronis[9], Rayfel Schneider[10], Grant Schulert[11], Richard Vehe[12], Karen Onel[13], for the SJIA workgroup of the CARRA JIA subcommittee

[1]Seattle Children's Hospital, University of Washington School of Medicine, Seattle, WA, USA

[2]Children's Mercy-Kansas City, University of Missouri-Kansas City, Kansas City, MO, USA

[3]Comminuty stakeholder CARRA member

[4]Texas Children's Hospital, Baylor College of Medicine, Houston, TX, USA

[5]McGovern Medical School at UT Health, Houston, TX, USA

[6]Boston Children's Hospital, Harvard Medical School, Boston, MA, USA

[7]Rutgers Robert Wood Johnson Medical School, New Brunswick, NJ, USA

[8]Mayo Clinic, Rochester, MN, USA

[9]Children's National Health System, The George Washington School of Medicine, Washington, DC, USA

[10]The Hospital for Sick Children, University of Toronto, Toronto, ON, Canada

[11]Cincinnati Children's Hospital, University of Cincinnati College of Medicine, Cincinnati, OH, USA

[12]Masonic Children's Hospital, University of Minnesota, Minneapolis, MN, USA

[13]Hospital for Special Surgery, Weill Cornell Medical College, New York, NY, USA

Background:

There has been rapid development in new treatments for systemic juvenile idiopathic arthritis (SJIA) but there is no consensus on when and how to withdraw medications. There are validated criteria for clinical inactive disease (CID) in juvenile idiopathic arthritis (JIA); however, as SJIA is considered an autoinflammatory disease there may be additional criteria to consider. The SJIA CARRA workgroup sought to obtain opinions on CID in SJIA and how physicians approach withdrawal of medications.

Methods:

An anonymous electronic survey using REDCap was sent to 100 randomly selected voting members of CARRA. The survey elicited physicians' opinions on CID in SJIA and physicians' current approaches to withdrawal of medications in CID. Descriptive statistics were used to analyze the data.

Results:

Eighty-three of the 100 surveyed CARRA members completed the survey including 7 members that opted out, as they are not involved in clinical care of SJIA. The vast majority of participants (88%) agreed with the current criteria for CID in SJIA. Stated reasons for dissent included lack of ferritin and other inflammatory markers in the criteria, presence of uveitis in the criteria, preference for extending duration of morning stiffness, preference for changing duration of time required for CID, imprecision of physician global score and lack of patient/parent-reported outcomes. Ninety-three percent agreed with the current definition for clinical remission on medications (CRM) in SJIA. Disagreement was due to preference for 1 year (not 6 months) of inactive disease to meet CRM. Most felt it was necessary to meet CRM (78%) before tapering medications other than steroids, but others stated preferences for withdrawing other therapy before 6 months of CID. Most members (76%) reported using the CARRA SJIA consensus treatment plans always or the majority of the time. All members reported that they weaned steroids first in SJIA patients on combination therapy, 47% reported waiting greater than 6 months before tapering additional medications, and 37% preferred waiting only 2-6 months. An equal number of members (35% each) reported tapering methotrexate over more than 6 months and 2-6 months; however a higher proportion (39%) preferred tapering anakinra, canakinumab and tocilizumab more quickly over 2-6 months and favor spacing the dosing interval for canakinumab and tocilizumab. When patients are on combination therapy with methotrexate and biologics, 58% preferred tapering methotrexate first while most others would consider patient/family preference and adverse effects to guide their choice.

Conclusion:

Most CARRA members surveyed are using previously published consensus treatment plans for SJIA and agreed with validated definitions of CID and CRM. There was also agreement with tapering steroids first in SJIA, but there was considerable variability with all other medications. Further work will need to be done to develop consensus in withdrawal plans for medications in SJIA.

The study was approved by Seattle Children's Hospital's Internal Review Board, ID: STUDY00000532, and Hospital for Special Surgery's Internal Review Board, ID: 2017-0276.