LUNG COMPLICATIONS IN SJIA

Lung disease has not been typically associated with SJIA, but recently, a small number of patients have been diagnosed with severe lung disease: Interstitial Lung Disease & Pulmonary Alveolar Proteinosis. Some of these patients also have a history of recurrent MAS.

Some patients have rapid escalation of lung symptoms, while for others, it has proceeded slowly or stabilized.

Lung complications are serious, so please discuss with your rheumatologist if you notice any symptoms.

The first signs of lung issues in SJIA can be:

-Swelling at edge of fingers or toes (or clubbing)

-Persistent, unexplained cough

-Breathlessness

EXPERT CONSULTS FOR REFRACTORY PATIENTS

Many SJIA kids do well with currently available medications. For kids with refractory disease, repeat MAS, and/or lung complications, it can be helpful to consult a specialist in SJIA/MAS who is more familiar with newer drugs.

This leaflet was reviewed by physicians who are experts in SJIA & MAS.

ABOUT THE SYSTEMIC JIA FOUNDATION

The SJIA Foundation is a patient-driven 501(c)(3) non-profit organization whose mission is to find a cure for Systemic JIA or Still's Disease.

The Foundation was started in 2014 by parents of a child diagnosed with Systemic JIA, MAS & Interstitial Lung Disease to help find a cure. It is run by parents & patients and collaborates with researchers across the world.

With your help, we support research, connect families, raise awareness & funds to find better treatment options and ultimately a cure.

We are stronger together! Join us.

SCAN FOR PATIENT RESOURCES

Our patient page includes:

- Online community for patients & caregivers
- Events organized by Systemic JIA Foundation
- How you can get involved
- Patient support groups

www.systemicjia.org/patients

PATIENT SUPPORT GROUPS ON FACEBOOK

Some of the bigger patient groups listed below. You can search for more.

- **SJIA Parents Network (excellent support group only for parents & guardians)** www.facebook.com/groups/SJIAParents
- **International Still's Disease Support Center (for parents & patients)** www.facebook.com/groups/StillsDisease
- **Adult Onset Still's Disease Support Group (for parents & patients)** www.facebook.com/groups/22056595992

Systemic Juvenile Idiopathic Arthritis (SJIA) or Still's Disease

WHAT IS STILL'S DISEASE OR SJIA?

- Systemic Juvenile Idiopathic Arthritis (SJIA), also known as Still's Disease, is a rare immune disease of unknown etiology (i.e., we don't understand what causes this disease).
- While pediatric SJIA is considered a JIA (Juvenile Idiopathic Arthritis) subtype, it differs significantly in symptoms and treatments from other JIAs. It's considered an autoinflammatory condition, though not monogenic in origin.
- The adult form of SJIA is Still's Disease (Adult Onset Still's Disease).
- The disease presents in three forms: (a) Monocyclic: one episode with no recurrence, (b) Polycyclic: active episodes with quiet periods between, and (c) Persistent: chronic disease. Approximately 30-50% of children experience Monocyclic disease, which resolves either naturally or with medication.
- There is no cure for SJIA. However, disease can go into remission, either on or off medications.

Labs to Watch with SJIA

- **CRP** (High)
- **ESR/Sed Rate** (High)
- **Ferritin** (High)
- Platelets (High)
- Hemoglobin (Low)
- White Blood Cells (High)
- **ALT** & **AST** (High)
- Speciality Labs: IL-18

SJIA SYMPTOMS

- Recurrent Fevers
- Salmon-pink Rash
- Arthritis (swelling of joints)
- Pain
- Enlarged lymph nodes
- Enlarged liver and/or spleen
- Serositis, pericarditis
- Myalgia (muscle pains)
- Pleurisy, pleural effusions
- MAS (Macrophage Activation Syndrome)

HOW IS SJIA TREATED

Commonly Used Drugs

- **NSAIDs** (Nonsteroidal anti-inflammatory drug; ibuprofen, naproxen, meloxicam etc.)
- **Oral Corticosteroids** (prednisolone)
- **IL-1 Blocking Biologics**: anakinra (Kineret), canakinumab (Ilaris), rilonacept (Arcalyst)
- **IL-6 Blocking Biologics**: tocilizumab (Actemra), and sarilumab (Kevzara)
- **Methotrexate**
- **Jak Inhibitors** such as: tofacitinib (Xeljanz), baricitinib (Olumiant), upadacitinib (Rinvoq), and ruxolitinib (Jakafi)
- Some new targeted drugs for blocking IL-18 are in clinical trials in USA & Europe.

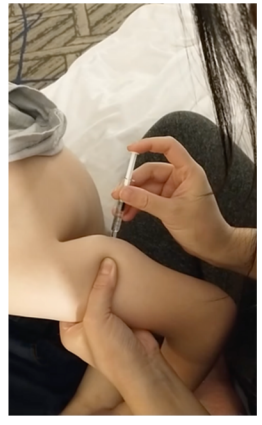
Refractory SJIA

- When commonly used drugs do not control disease, it is called refractory SJIA, which can be characterized by uncontrolled arthritis, and/or systemic disease, recurrent MAS and/or lung disease.
- For refractory cases, multiple drugs might be used, or other immunosuppressives are added.
- It is important to find the right combination since uncontrolled SJIA is associated with recurrent episodes of MAS and other complications like lung disease.

WHAT IS THE OUTCOME

There is no cure for SJIA, and the disease course varies from child to child. For many patients, the disease goes into remission on its own or aided by medicines. There is a subset of refractory patients with hard-to-control disease, with multiple MAS episodes, lung complications, and/or uncontrolled arthritis. There is about a 5-10% mortality rate with each case of MAS.

MANAGING YOUR SJIA CHILD: TIPS FROM PARENTS



- The most important thing is to find a good rheumatologist, ideally one with familiarity with SJIA, given that it is a rare disease.
- The thermometer is your friend. Buy a good one, and use it so you know what your child's normal is.
- Take pictures of new rashes so you can show them to your doctor or compare them with previous ones.
- Some SJIA meds are daily/weekly shots that the parents administer. Administering shots is always tough, but it does get easier over time.
- Learn to track your child's inflammatory labs (e.g., CRP, ESR, Ferritin, Platelet, WBC, Hemoglobin, AST, ALT). Get a copy of labs from your doctor/center or obtain it from online health record systems, e.g., MyChart.
- Some parents, especially those with lung complications, have bought a finger pulse oximeter (available on Amazon.com or drug store) for tracking heart rate and oxygenation for their child.

MACROPHAGE ACTIVATION SYNDROME (MAS)

MAS is a life-threatening complication of SJIA

- Over 20% of SJIA patients present initially with MAS. For many patients, this is the only episode of MAS. Other patients can suffer from recurrent MAS episodes.
- Common symptoms of MAS are: (a) high persistent fever (unlike fevers with SJIA which come and go), (b) irritability, (c) lethargy/confusion, and (d) headache.
- Early diagnosis of MAS can be based on lab patterns rather than clinical symptoms. For those at risk for MAS, even a minor illness with fever should warrant the need for lab tests.
- MAS can also be subclinical (meaning only seen in lab patterns).
- Biologic drugs can mask labs making MAS harder to diagnose.
- Escalating MAS may cause seizures, coma, organ failure, and even death.
- **MAS is treated with IV Pulse steroids. Some centers use high doses of anakinra or add Cyclosporine.**
- Treating severe cases of MAS is challenging and it is important to begin treatment early.
- A new targeted drug for MAS called emapalumab is in clinical trials in the USA & Europe.

Labs to Watch with MAS

- **CRP** (High)
- **ESR / Sed Rate** (Low)
- **Ferritin*** (V. High)
- **Platelets** (Low)
- **Hemoglobin** (Low)
- **White Blood Cells** (Low)
- **ALT & AST** (High)
- Speciality Labs: **IL-18**, **CXCL9**

** Ferritin is the most reliable biomarker of MAS*

MAS in SJIA Criteria (from PReS/EULAR 2016)

Fever and Ferritin >684 ng/ml plus any TWO of the following:

- Platelets ≤181 x 10^9/L,
- AST >48 u/L
- Triglycerides >156 mg/dL and/or Fibrinogen ≤360 mg/dL