Indian Academy of Pediatrics (IAP)

GUIDELINES FOR PARENTS

Rheumatic Disorders

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10 FAQs on RHEUMATIC DISORDERS

1. I have heard of rheumatic disorders in adults but can they affect children too! What are the red flag signs to suspect and seek a doctor's opinion?
2. My child is having pain in both legs during late evening often relieved with massage. Is it a sign of evolving serious rheumatic disorder?
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10. My daughter is diagnosed with lupus at 9 years of age. Our family is distressed about the life-threatening nature of the disease! What should we do to improve her quality of life and would she be able to conceive during adulthood?

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Rheumatic Disorders

Q1

I have heard of rheumatic disorders in adults but can they affect children too! What are the red flag signs to suspect and seek a doctor's opinion?

Rheumatological (pronounced "RHEUMA—TO-LOGICAL")/rheumatic diseases are autoimmune and inflammatory diseases that cause our immune system to attack various joints, muscles, bones, blood vessels, and other organs. Often thought to affect only the elderly, these diseases have definite and specific afflictions to the pediatric age group (0–18 years). Doctors who care for such disorders in children are termed as "pediatric rheumatologists".

Inflammatory conditions commonly seen include juvenile idiopathic arthritis (JIA), systemic lupus erythematosus (SLE), juvenile dermatomyositis (JDM), Kawasaki disease (KD), Henoch–Schönlein purpura (HSP), etc. In addition, various non-inflammatory conditions such as mechanical pains (growing pains), benign joint hypermobility syndrome, pain amplification syndromes besides bony pains due to periodic growth spurts (Sever's disease, Osgood–Schlatter disease, etc.) can have musculoskeletal symptoms.

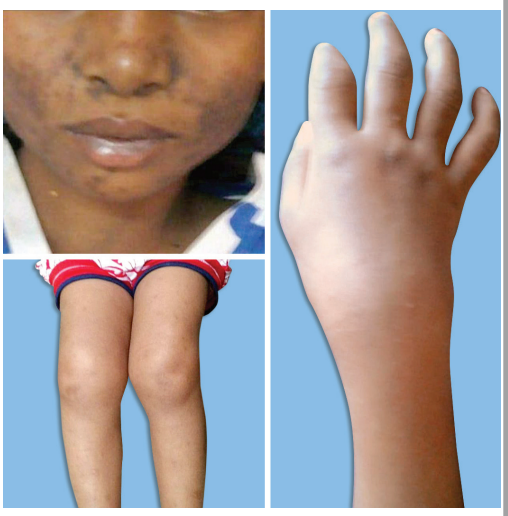

Fig. 1: Reflecting rashes and joint swelling
Courtesy: Dr Vijay Viswanathan.

The common signs and symptoms that may herald the onset of such conditions include persistent/unexplainable limps (specific to morning hours), fever, rashes, redness of eyes, mouth ulcers, joint swelling, nonhealing ulcers, and muscle weakness **(Fig. 1)**. Many times, nonspecific symptoms such as weight loss, tiredness, arrest/regression of motor milestones are noted at onset especially in younger children. The first consultation usually for any of these complaints should be to a general pediatrician who would refer to a pediatric rheumatologist as needed.

Q2

My child is having pain in both legs during late evening often relieved with massage. Is it a sign of evolving serious rheumatic disorder?

Benign (not serious) nocturnal (night-time) pain of childhood (often referred as growing pains) is a common problem which usually begins in the age groups of 2–6 years. The process seems to arise from children being extremely flexible ("Loose jointed") with excessive ligament laxity. Such children usually have a low muscle tone. This coupled with lower bone strength results in overusage of the ligaments throughout the day causing night-time pains. It often gets relieved with massage/warm fomentation.

It may cause more anxiety in families, but most often children outgrow the pains with age. It generally does not evolve into a serious disorder.

However, night-time pains which get worsened by massage or pressure, associated with fevers, disturbed sleep may be a warning sign and needs to be assessed by a specialist for possible sinister conditions.

Q3

What causes these diseases? Do these disorders run in the family or pose risk to the sibling?

We are protected from infections caused by several bacteria and virus by our immune system. Our immune system can differentiate between what has to be destroyed being potentially harmful and foreign and what is ours. In most of the rheumatic disorders, e.g., juvenile idiopathic arthritis, juvenile dermatomyositis, systemic lupus erythematosus, our immune system loses its ability to identify "self" from "foreign" cells and attacks our own body thus known as autoimmune diseases [immune system working against self (auto)]. There may be interplay between the host (affected child) and the environment, e.g., an untreated sore throat by specific bacterial species of streptococci can trigger an abnormal immune response resulting in rheumatic fever. Antibiotics can treat the throat infection, cease the immune system activity, and prevent further attacks. However, most of the time the underlying trigger for autoimmunity remain unknown.

These are not hereditary diseases as they are not transmitted directly to the children from their parents. However, there are genetic factors that predispose to the disease. It is emphasized that, they are *not* predestined to get autoimmune diseases. There is no genetic testing or prenatal diagnosis for most of these illnesses. There is an agreement that the cause may be a combination of exposure to environmental trigger in presence of genetic predisposition. However, two children even twins are rarely affected with the same disease. It is also important to note that these diseases are neither infectious nor contagious.

Q4

Can I continue the vaccination schedule of my child with underlying rheumatic disorder?

Vaccination schedule should be discussed with your doctor from the first visit onward. It is ideal to have your child vaccinated up-to-date. If your child is on medicines that suppress immunity like high-dose steroids and biologics, live-attenuated vaccines should preferably be avoided; however, MMR vaccination maybe considered. Vaccination should preferably be administered during quiescent stage of the underlying disease. This is because there is a potential risk of introducing these infections due to the decreased immune defense system. OPV is also contraindicated in family members living in houses of children who are on immunosuppressive therapy. Live vaccinations must be postponed for at least 6–9 months after receiving intravenous immunoglobulins.

Vaccines that have infectious proteins and no live microorganisms [e.g., Diphtheria, Tetanus, and Pertussis (DPT), injectable polio, hepatitis B, pneumococcus, hemophilus B; meningococcus, influenza, and human papilloma virus) can be given safely during the course of treatment. Vaccine may give reduced protection due to suppression of immunity. Pathogen-specific antibodies could be measured to assess vaccine response and need for revaccination after consultation with your physician.

The present recommendation is that the vaccination schedule for nonlive composite vaccines is followed. The only exception is children with severe active illness who must not receive any vaccinations until the disease is controlled.

Q5

Are they curable and how long to continue treatment?

Many of this illness are chronic, characterized by remission and relapses. Timely diagnosis and initiation of treatment is the first target towards remission. Compliance to the management protocols, ensuring continuation of all activities of daily living, psychological support, and coping skills by the family also aid significantly in early remission.

The duration of treatment varies for each child according to the underlying disease and severity. It may be as short as 6 weeks for a child with typical uncomplicated Kawasaki disease to lifelong for a child with multiorgan involvement due to SLE. The therapy should last as long as the underlying disease persists. Most children with SLE need long-term low-dose steroids to keep the disease under control and prevention of flares. Most children with illness such as polyarticular JIA, systemic JIA, and JDM need treatment for 24–36 months, however, some may need therapy for several years. Treatment may be reduced gradually over several months, after the child has inactive (clinically and normal blood tests) disease. All children with rheumatic disorders must stay in long-term follow-up even if they have sustained remission and should be transitioned to the adult physician/rheumatologist in late adolescence.

Q6

What are the available treatment options for management of common rheumatic diseases? What should I monitor till we get back to you for follow-up?

A wide range of treatment options are available for management of rheumatic disorders as mentioned here:

- *Drugs*: Nonsteroidal anti-inflammatory drugs (NSAIDs) (e.g., naproxen), steroids (given either systemically or as joint injection), disease modifying antirheumatic drugs (DMARDs) (e.g., methotrexate), and biological agents (e.g., etanercept, adalimumab) are the mainstay of treatment.
- *Physiotherapy*: For management of pain, restoration of normal mobility and prevention of deformity is equally important. Encourage sport activities such as swimming, cycling or yoga as feasible.
- *Corrective surgery* (for severe deformity and limb-length discrepancy) may be required in few cases after skeletal maturity.

Choice of drugs depends on type and severity of disease. While many oligoarticular JIA children can be managed successfully with NSAIDs and joint injections, polyarticular JIA and systemic onset JIA most often require early aggressive treatment with DMARDs and targeted agents called "biologicals" wherever needed. While NSAIDs and DMARDs are inexpensive, targeted treatment with biological agents are relatively expensive. However, more often in such situations, the benefits overweigh the risk of progressive disease.

There is an increased risk of acquiring infections such as tuberculosis, with use of immunosuppressive drugs particularly biological agents. It is important to rule out tuberculosis prior to starting treatment and monitor for the symptoms of same throughout. The risk of infection can be curtailed by maintaining a symptom diary, regular follow-up and vaccination as advised by your treating physician.

Q7

Are there any dietary restrictions for my child with underlying rheumatic disorder? Does the disease affects the growth and development of my child?

As such, no dietary restrictions are required for rheumatic disorders of childhood. Since these are chronic inflammatory disorders, growth retardation and malnutrition are seen in these children. So, a balanced healthy diet having recommended proportion of fat, and carbohydrate with enough micronutrients is necessary for them.

The rheumatic disorders may affect normal growth of the child particularly in sJIA, polyarticular RF positive JIA, SLE and others due to disease-related inflammatory process. Growth abnormalities in these children may also be contributed by the use of steroids. So monitoring of growth parameters is important. You should take your child for regular follow-up for monitoring of growth and development. With proper management and supportive treatment most of them will not have significant growth problem.

Q8

My son is having JIA; would it affect his schooling and he is quite stressed about his participation in sport activities and ability to live normal life! Does this affect life expectancy?

Juvenile idiopathic arthritis does not affect cognition and these children must attend regular schools, unless limited by disease flare. The parents should educate teachers and suggest the following measures to help their child:

- Sitting at sides of the classroom allows them walking/changing posture to mitigate the stiffness experienced by them due to prolonged sitting.
- Engaging them in activities which promote walking and relieve stiffness such as collecting papers, monitoring, etc.
- Two set of books (if feasible) may reduce additional burden of weight bearing.
- Allowing them for comfortable position for instillation of eye drops (if required for associated uveitis).
- Use "fat" pen/pencil **(Fig. 2)** and felt-tip pen and stretching of hands every 10–15 minutes to provide ease while writing.
- Supervise the medication administration during school hours.
- Vocational training during secondary schooling for those with significant disability.
- Physiotherapy and exercises should enhance their play and leisure activity.

The disability because of JIA will depend on the underlying subtype; systemic and polyarticular forms may be more disabling than oligoarticular JIA. Approximately 30% of children with enthesitis-related arthritis experience limited spine mobility and 40–50% will have ongoing disease activity as they transition into adults and a few can require joint replacement (<5%).

The mortality in JIA is <1% and is mainly due to life-threatening complications such as macrophage activation syndrome and infections associated with steroid use. However, with the availability of more aggressive therapeutic regimens, biologicals and close supervision the long-term outcome is optimistic for JIA.

Fig. 2: Fat pencil/pen.

Q9

Giving weekly methotrexate is becoming a real challenge for me. He feels nauseating throughout the day and also pukes at times. Please help me out with this!

Nausea, abdominal discomfort, and gastrointestinal symptoms are the most commonly associated side effects of methotrexate. However, this is a very effective and safe drug among the available arsenal of various disease modifying anti-rheumatic drugs for controling arthritis, and this should encourage you to comply with the prescribed dosing regimen.

To reduce these side effects, you must use folic acid supplementation as prescribed on a day other than on which you are taking methotrexate. Other options to reduce the side effects are:

- Shifting to a subcutaneous route after consultation with your physician can also help in attenuating these side effects. You/your ward can learn administering methotrexate through a subcutaneous route **(Fig. 3)**; self-learning shall save your time and ensure compliance.
- If the symptoms are too bothersome, taking antiemetics (ondansetron) on the day of methotrexate after consultation with your doctor shall give respite.

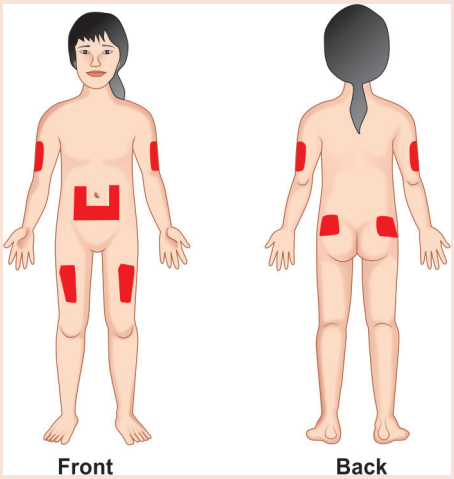

Fig. 3: Sites for methotrexate subcutaneous injections.

Acknowledgment: Mr Rajesh Saini, CMET, AIIMS, New Delhi for helping in preparing the representative Figures 2 and 3.

Q10

My daughter is diagnosed with lupus at 9 years of age. Our family is distressed about the life-threatening nature of the disease! What should we do to improve her quality of life and would she be able to conceive during adulthood?

Childhood lupus is a multisystem disease which can affect almost every organ of the human body. The most common cause of mortality being kidney or cardiopulmonary involvement and infections. Over the past few years, with the availability of strong immunosuppressive drugs their life expectancy is nearly (>95%) normal.

Regular follow-up, maintaining a symptom diary including blood and urine tests, and compliance to drugs are the most important keys for defeating the disease related damage. In addition your child needs evaluation for steroid-related side-effects (weight gain, eye changes, weakness of bones, etc.) and you must *avoid self-administration* of these drugs:

Disease flares can be prevented by adhering to the following:

- Sun-protection (sun-screen and protective clothing)
- Avoid potential source of infections: wear facemask, vaccination
- Age appropriate physical activity, exercises and vitamin D intake
- No to active/passive smoking
- Seek medical advice whenever your child experiences fever, oral ulcer/skin rash, increased hair fall, etc.

These children may have impaired reproductive functions due to the disease and the gonadotoxic drugs such as cyclophosphamide. However, the prepubertal gonads are relatively protected from ill-effects of gonadotoxic drugs. With the advent of better modalities of disease control the lupus-related pregnancy loss is <20% with majority of pregnancies being uneventful and normal fetomaternal outcome.