



Rheumatology, Cancer, and Gastrointestinal Disorders
Victory Junction Medical Staff and Volunteer Training



Objectives – What's the target?

- Rheumatology
 - Review basic pathophysiology of the most common rheumatic illnesses in children
 - Discuss common complications and emergencies
 - Discuss treatment of rheumatic disorders in children
 - Discuss considerations for campers
- Cancer
 - Review basic pathophysiology of the most common childhood cancers
 - Discuss common complications and emergencies
 - Discuss treatment of childhood cancer and complications
 - Discuss considerations for campers
- GI Disorders
 - Review basic pathophysiology of the most common GI disorders in children
 - Discuss common complications and emergencies
 - Discuss treatment of selected GI disorders
 - Discuss considerations for campers



The target is camper safety! Better understanding of the diseases our campers have means better care!

Rheumatology

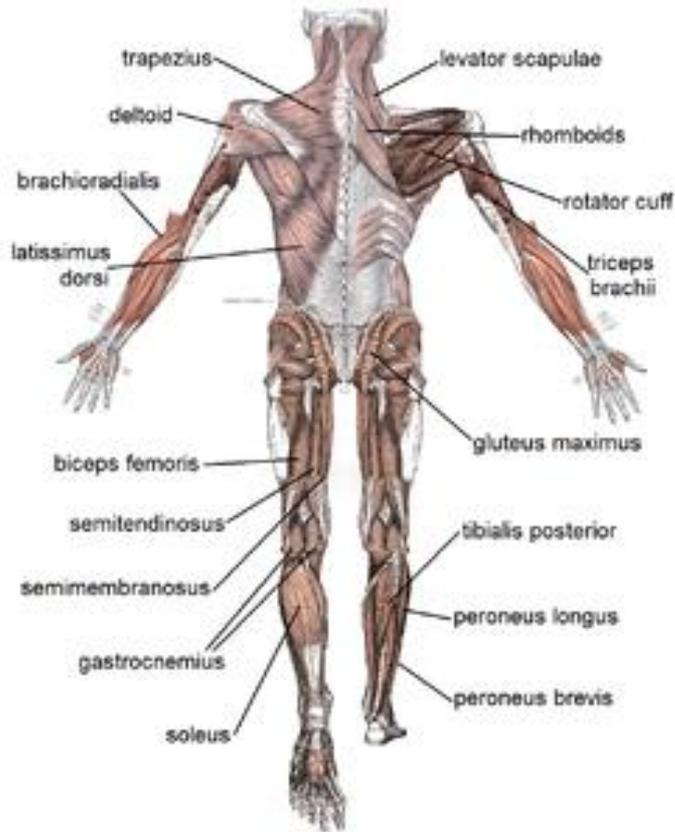


Rheumatology

Rheumatic illnesses are those of the **muscles, joints, and connective tissue.**

Juvenile arthritis is the most common chronic rheumatologic disease in children.

About 294,000 children < age 18 in the U.S. have arthritis or another rheumatic condition.



Juvenile arthritis is usually an autoimmune illness.

The previously used term “Juvenile rheumatoid arthritis” has been broadened to “Juvenile idiopathic arthritis.”

Rheumatology – Juvenile Arthritis

Juvenile idiopathic arthritis (JIA) is the most common juvenile arthritis and encompasses several types of arthritis, the most severe of which is **systemic JIA**.



What causes JIA?

- A flaw in the immune system causes inflammation (cytokine release) that causes break down of the cartilage and bone, and also activates the release of rheumatoid factor, which is responsible for the degradation of joint tissue. JIA is an **autoimmune** disease.



What are symptoms of systemic JIA?

- **Pain, stiffness, and swelling** in predominantly large joints
- Generally affects one side of the body
- Mild **fever** at onset or flare of illness



How is JIA diagnosed?

- Symptoms of arthritis present for > 6 weeks in child < 16 years
- Serological tests for rheumatoid factor (however, seldom detected)
- **Diagnosis of exclusion**

Rheumatology – Juvenile Arthritis

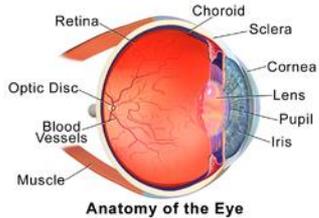
How is systemic JIA treated?

Treatment depends on severity at initial presentation, and may be adjusted over time based on response to previous treatments and continued evaluation of severity. Severity is scored by the number of affected joints and presence or absence of active systemic features. Commonly used medications include:

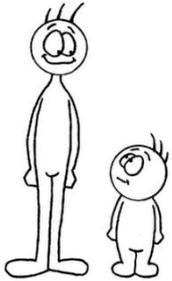
- NSAIDs (i.e. ibuprofen, naproxen sodium)
 - Reduce inflammation
 - Given as oral medication
- Methotrexate
 - Disease-modifying anti-inflammatory medication, type of chemotherapy/biotherapy
 - Should be taken with folic acid
 - Given as oral or subcutaneous medication
- Anakinra
 - Inhibits an inflammatory cytokine (IL-1) responsible for the pathogenesis of JIA
 - Used with or without steroid therapy
 - Usually given subcutaneously
- Intra-articular glucocorticoid injection
 - Delivers anti-inflammatory steroid directly to joint
- Tocilizumab
 - Inhibits an inflammatory cytokine (IL-6) responsible for the pathogenesis of JIA
 - Usually a periodic IV infusion

Rheumatology – Juvenile Arthritis

What are common complications for children with JIA?



Uveitis – inflammation inside the eye. This may cause pain, vision changes, redness, and/or sensitivity to light.



Growth problems – Children with JIA may not grow as quickly as their peers. They may also have asymmetric bone growth or joint contractures due to inflammation.



Immunosuppression – many medications to treat JIA suppress the immune system and increase risk of infection. Fever is a big deal and should be addressed immediately!

Flare-ups are the most severe complication of JIA. The illness can go into remission, when the child has no symptoms, and then have recurrent episodes of “flaring up.”



Rheumatology – Juvenile Arthritis

Helpful Medical Check-in Questions:

- When was your last flare-up? What are the early warning signs for you?
- Is there anything you do to prevent worsening pain? (Some campers use a wheelchair for long distances, or other mobility aids.)
- How do you treat joint pain? (Review any PRN medications and Forbidden OTCs)
- Do you have any target joints?
- Is there anything else I should know to make sure you have a safe and fun week at camp?

Update “medical check in” and “unit nurse” notes in CampSite as applicable.



Rheumatology – Ehler-Danlos

Ehler-Danlos syndrome (EDS) is a group of connective tissue disorders, resulting in increased flexibility and fragility of soft connective tissues. There are 6 types of EDS, based on genetic defect and resulting effects, although symptoms of each type may vary in individuals:

Classical type (types I & II): Occasional internal organ fragility, autosomal dominant or recessive

Hypermobility type (type III): Most common type, autosomal dominant inheritance

Vascular type (type IV): Fragility of arteries, intestines, and other organs, particularly serious type

Kyphoscoliosis (type V): Kyphoscoliosis, fragility of arteries and eyes. Very rare, autosomal recessive

Arthrochalasia (type VII, A & B): Very rare, autosomal dominant

Dermatosparaxis (type VII C): Very rare

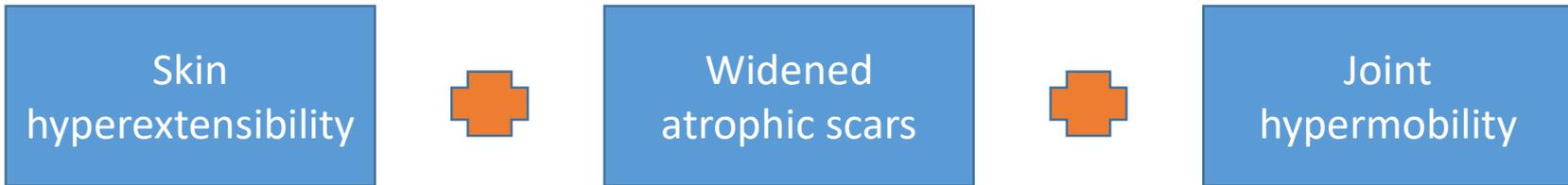
EDS is an inherited disorder, and each type “runs true” in a family. EDS affects all races, ethnicities, and genders.



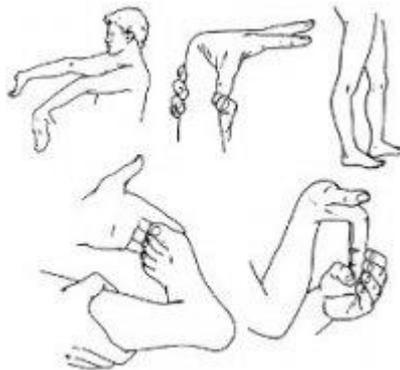
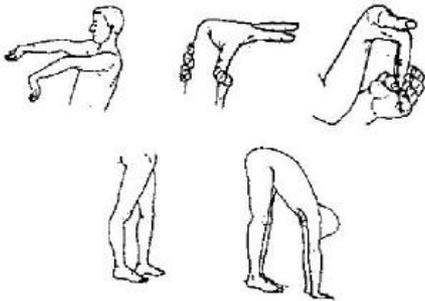
Rheumatology – Ehler-Danlos

The clinical spectrum of EDS ranges from mild skin and joint hyperlaxity, to severe physical disability and even life threatening vascular complications.

There are 3 major diagnostic criteria for the *classic type EDS*:



Skin often feels **velvety and smooth** to touch, and will easily stretch but snap back quickly.



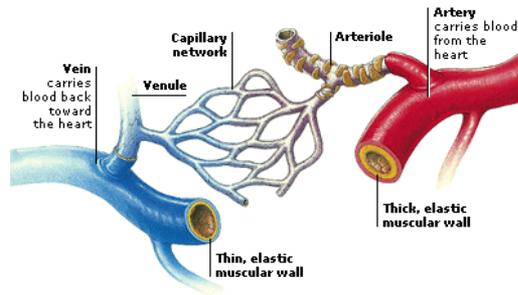
Joint hypermobility is usually generalized, and can cause many complications including:

- Hip dislocation at birth
- Congenital club foot
- Joint effusions
- Repetitive joint dislocation (spontaneously or with minor trauma)
- Chronic and recurrent pain
- Premature osteoarthritis

Rheumatology – Ehler-Danlos

EDS Hypermobility type

- **Most common type**, though difficult to diagnose
 - Genetic basis largely unknown
 - Difficult to differentiate from benign joint hypermobility syndrome
- Presentation: **generalized joint hypermobility** and **skin manifestations**
 - Usually more subtle than in classic type EDS



EDS Vascular type

- Worst prognosis
 - Risk of spontaneous arterial rupture, usually in 3rd or 4th decade of life
- Presentation: initial presentation is often **excessive bruising**, also **thin, translucent skin**, and **joint hypermobility limited to the hands**

Rheumatology – Ehler-Danlos

Precautions for campers with Ehler-Danlos may include:

- Avoiding contact sports, and wearing protective pads to avoid excessive bruising and hematomas
- May prefer to use acetaminophen over NSAIDs for pain due to NSAID effect on platelet function
- May have chronic or recurrent musculoskeletal pain
- Be aware of risk for joint dislocation spontaneously or with even minor trauma
- Be aware of risk for increased scarring with any cutaneous injury

Check “Forbidden OTCs” in
CampSite!

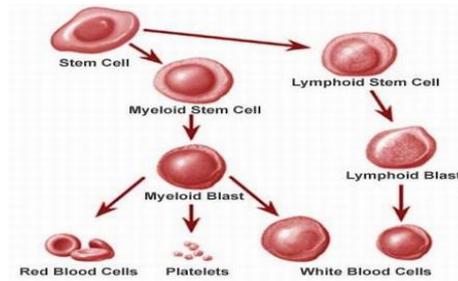


Childhood Cancer



Cancer

- About 10,380 children under age 15 will be diagnosed with cancer in 2016
- About 1,250 children under age 15 are expected to die from cancer in 2016
- Cancer is the 2nd leading cause of death in children, behind accidents
- Overall 5 year survival rates are about 80%
- Leukemias, cancers of the bone marrow and blood cells, are the most common childhood cancer (about 30% of all childhood cancers)



- Brain and central nervous system tumors are the second most common childhood cancer (about 26% of all childhood cancers)



Cancer – Types of childhood cancer

- **Leukemia** – Cancer of the bone marrow cells that become white blood cells. Acute lymphocytic leukemia (ALL) of the B or T cells and acute myeloid leukemia (AML) are common types. Pre-B cell ALL is the most common and has the highest survival rate (for children with standard risk profile ALL, diagnosed between ages 2 and 10). AML is much higher risk. Treated with chemotherapy, and in some cases stem cell transplant.
- **Brain/CNS tumors** – Solid tumor, many different types with prognosis and symptoms depending on location, size, and type of tumor. Often treated with surgery, chemotherapy, and radiation (or in some cases high dose chemotherapy with stem cell rescue, called an autologous stem cell or bone marrow transplant).
- **Lymphomas** – Cancer of the lymph cells, classified as either Hodgkin’s or non-Hodgkin’s, both types are more common in older children and teens. Treated with chemotherapy, radiation, and in some recurrent cases stem cell transplant.
- **Neuroblastoma** – Solid tumor of the nerve cells, often presents as abdominal mass, most often diagnosed in infants and toddlers and often advanced stage at diagnosis. Treated with chemotherapy, surgery, radiation, stem cell transplant, and immunotherapy.
- **Wilm’s tumor** – Solid tumor of one or both kidneys, most often diagnosed in children ages 3-5, usually treated with chemotherapy, surgery, and radiation.
- **Osteosarcoma/Ewing’s sarcoma** – Both are primary bone tumors and occur most often in teens. Treatments include chemotherapy, surgery, and radiation (in Ewing’s only).
- **Rhabdomyosarcoma** – Most common soft tissue tumor in children, can occur anywhere there is skeletal muscle. Treated with chemotherapy and radiation.
- **Retinoblastoma** – Cancer of the retina, usually diagnosed before age 2. Retinoblastoma has a strong genetic component, and is most often treated by surgically removing the eye with or without chemotherapy.

Cancer – Childhood cancer treatments

Type of Treatment	Chemotherapy	Radiation	Stem Cell Transplant (Bone Marrow Transplant)	Immunotherapy
How does it work?	Many different classes, all disrupt the cell cycle to prevent cancer cells from multiplying. Chemotherapy has a systemic effect, and affects all rapidly dividing cells (cancer cells, epithelial cells, etc.). Can be given as oral, intravenous, and/or intrathecal medication.	Primarily used with solid tumors, radiation is targeted at tumor site to kill cells by damaging their DNA	Deplete the immune system with very high dose chemotherapy \pm radiation, and rescue with the person's own stem cells (auto) or healthy immune cells from a donor (allo). Leukemia and bone marrow diseases require an allogenic transplant, solid tumors often treated with autologous stem cell rescue.	Newer therapies and focus of many clinical trials, utilize the body's immune system to fight cancer cells.
Acute side effects	Nausea, vomiting, fatigue, anemia, neutropenia, thrombocytopenia, mucositis, increased risk of infection	Nausea, vomiting, fatigue, anemia, mucositis or skin burns, sensitive skin, increased risk of infection	Similar to chemotherapy and radiation, higher doses result in more severe side effects. SCT is a life threatening process, children are often inpatient for at least one month.	Varies with type, may include systemic inflammatory response, nausea, anemia, liver and kidney involvement
Long term side effects	Varies widely, may include hearing loss, neuropathies, cardiac and respiratory effects, GI/liver/kidney dysfunction, reproductive dysfunctions, and secondary malignancy	Decreased growth of bones post exposure to radiation (i.e. short stature, craniofacial changes), skin and gut (epithelial) changes, reproductive dysfunction, secondary malignancy	Graft-versus-host-disease can be an acute and/or chronic complication, and may involve the skin, GI system, or liver. Cumulative long term effects of chemo and radiation.	Newer treatments, long term effects are not yet well documented.

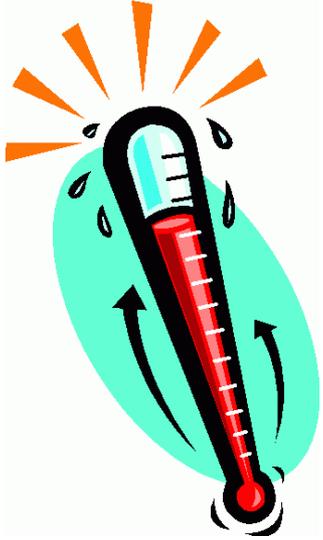


Childhood cancer – Common complications and management

Complication	Cause	Treatment
Neutropenia (decreased white blood cell [including neutrophil] count = Increased risk of infection)	Chemo and radiation target rapidly dividing cells, including white blood cells. 7-10 days after chemo is the nadir, or lowest point in the immune system.	Sometimes a medication is prescribed to stimulate recovery of white blood cells. Preventing infection and prompt treatment of fevers is critical. Some campers may take Bactrim or Septra (TMP/SMX) to prevent PCP pneumonia.
Anemia (decreased hemoglobin and hematocrit = fatigue, decreased stamina, headaches)	Chemo and radiation temporarily stop bone marrow production of red blood cells.	Encourage rest and hydration. If extremely low, treated with blood transfusion.
Bleeding (decreased platelet count = nosebleeds, easy bruising)	Chemo and radiation temporarily stop bone marrow production of platelets.	Bleeding precautions (no contact sports, soft toothbrush, etc.). At extreme lows, treated with platelet transfusion. Avoid NSAIDs.
Nausea/vomiting	Chemo and radiation affect the epithelial lining of the gut, causing nausea. Some medications activate a part of the brain that causes vomiting.	Hydration, rest. Medications may be prescribed, including ondansetron (Zofran), granisetron (Kytril), lorazepam (Ativan), diphenhydramine (Benadryl), or metoclopramide (Reglan)



Childhood cancer - Emergencies



Fever is an emergency in any child who is immunocompromised, such as those currently receiving treatment for cancer.

Fever = temp > 38 C (100.4 F)

If a camper with childhood cancer develops a fever, or any sign of infection, immediately notify the provider on call and the Director of Medical Operations.

Anticipate that the camper needs to be assessed for a source of infection, and that blood cultures and antibiotics may be ordered.



Cancer – Camper considerations

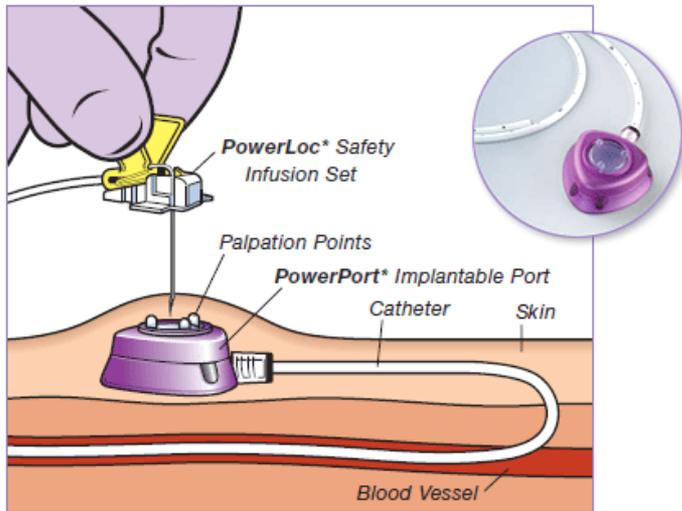
Precautions for campers with cancer or history of childhood cancer may include:

- **Fever precautions** (fever is treated as an emergency in any child who is immunocompromised or has a central line) 
- **Infection prevention** = handwashing, monitor for any open cuts/wounds/rashes
- **Zero tolerance for sunburn!** These campers may be extra sensitive to sunburn. 
- **Central line** maintenance  examine line, or Port site, at check-in, and make staff aware of line and location for activities (such as adventure). Cover any external line to protect from water, and change dressing immediately following swimming or showering.
- Offer a variety of food choices. Campers currently on treatment may prefer soft, bland foods, while some campers may prefer sour or spicy foods due to changes in their taste buds from chemotherapy.
- May prefer to **avoid NSAIDs**, due to effect of decreased platelet function. Check **Forbidden OTCs!**



Childhood cancer – Central Line care

There are 3 primary types of central venous access in children. These are used to infuse chemotherapy and other medications safely into a large vessel and draw blood samples, but can also increase risk for blood stream infections. Proper care is critical for the camper's health.



A port is an implanted venous access device. When not accessed, it lies beneath the skin. There is no activity restriction for campers with ports.

Assess the skin directly over the port for redness, swelling, pain, or any other signs of infection.



A PICC line (peripherally inserted venous catheter) is usually inserted through a vein in the upper arm and extends into the superior vena cava.

Assess the PICC dressing and insertion site. The PICC line and dressing must be protected during activities.



Central venous lines are tunneled catheters with 1-3 external lumens. There are not activity restrictions at camp, but staff should be notified of the line placement and avoid trauma to the site.

Assess the CVL dressing and insertion site, protect during activities, and change the dressing immediately after swimming or soaking in the shower.

Thank
you!



Childhood Cancer

Helpful questions for medical check-in:

- When was your last chemotherapy? (7-10 days out is when he will be most at risk for infection)
- Have you had any admissions recently? What for?
- When was your last “counts” check? (Any concern for bleeding risk or anemia during the week?)
- Where is your line? (Visualize and assess site.) Does it need to be flushed during your time at camp? When does the dressing need to be changed? Is there anything special we should know about your line?
- (If taking chemo while at camp) Do you take this on an empty stomach or with food? Is there anything else I should know about how you take this medicine?

Update the “medical check in” and “unit nurse” notes in CampSite as applicable.



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