The Immunocompromised Patient - Who Are They?

Recognizing and protecting the lesser-known immunocompromised population

By Heidi Moore, RN and Olivia Flynn, RN

Objectives

- Review statistics and pathogenesis of autoimmune diseases.
- Review most common pediatric autoimmune diseases.
- Review current treatment modalities with a special focus on biologic therapies.
- Review the school nurse role and responsibilities in caring for students receiving these medications.

Statistics

- AD affects 50 million Americans (>10%)
- 100+ known autoimmune diseases
- The prevalence is rising at alarming rates
- Females affected 75% more often than males
- Second highest cause of chronic illness
- Top cause of morbidity in women in the U.S.
**The Common Thread in AD**

- Genetic predisposition
- Presence of an environmental/hormonal trigger
  - virus, bacteria, medications, pollutants, hormones (pregnancy), stress
- HLA has been the most important single genetic trait in estimating susceptibility to autoimmune disease.

**HLA details**

- HLA has been a useful diagnostic/prognostic/management tool
- HLA reflects predisposition and not an inevitability of disease
- HLA B-27 found in 90% patients with ankylosing spondylitis
- HLA-DRB1 in rheumatoid arthritis may help predict severity and identify those patients that would benefit from earlier more aggressive therapy
- DNA technology now changing the use of HLA

**Immune system's complicated role**

Autoimmune reaction pathophysiology

Autoimmune Disease:
Immune signals heightened
Overview of Autoimmune Disease

- Autoimmunity persists and worsens
- Once damage occurs, categorized as AD
- Autoimmune disease can affect almost any organ of the body: heart, brain, nerves, muscles, skin, eyes, joints, lungs, kidneys, glands, the digestive tract and blood vessels

Do You Know the Disease?

- Joint pain, stiffness, fatigue, limp, fever
- Abdominal bloating, diarrhea, constipation, itchy skin rash
- Fatigue
- Increased thirst, increased urination, hunger, fatigue, blurred vision
- Rashes
- Rash, progressive muscle weakness, difficulty with position changes
- Dermatomyositis
- Hearing, preschool, episcleritis
- SLE
- Dry eyes, dry mouth, loss of sense of taste, weight loss, SOB
- Sjogren's
- Fever, weight loss, hair loss, mouth sores, fatigue, "butterfly" rash
- Lupus

Pediatric Autoimmune Disease

- Joint pain, stiffness, fatigue, limp, fever
- Abdominal bloating, diarrhea, constipation, itchy skin rash
- Fatigue
- Increased thirst, increased urination, hunger, fatigue, blurred vision
- Rashes
- Rash, progressive muscle weakness, difficulty with position changes
- Dermatomyositis
- Hearing, preschool, episcleritis
- SLE
- Dry eyes, dry mouth, loss of sense of taste, weight loss, SOB
- Sjogren's
- Fever, weight loss, hair loss, mouth sores, fatigue, "butterfly" rash
- Lupus
Pediatric Research

- Lack of specific specialty to treat autoimmune disease
- Lack of research funding
- Clinical trials vs. CARRA
- Medication use in children often not FDA approved

Clinical trials by pharmaceuticals vs. specialty-specific researchers
- Off-label use of medications
- Limited research on long term outcomes

Diagnosis in Pediatrics

- Clinical Symptoms
- Labs
- Imaging
- Scope

Rheumatological Autoimmune Diseases

Juvenile Idiopathic Arthritis (JIA, formerly JRA)
- Dermatomyositis
- Ankylosing Spondylitis
- Systemic Lupus Erythematosus (SLE)
- Uveitis
- Chronic lyme

Symptoms:
- Pain, joint swelling, stiffness, loss of motion
- Procr swallowing, muscle weakness, rash, SGB
- Back pain, swelling between vertebrae
- Affects any organ, butterfly rash, fatigue
- Blurred vision, redness of eye, light sensitivity
Autoimmune diseases of organs

- Celiac
- Crohn's Disease
- Ulcerative Colitis
- Type 1 Diabetes
- Congenital Heart Block
- Kawasaki

**Symptoms:**
- Gluten damages small intestine - diarrhea, abd pain
- Severe and persistent inflammation of GI tract
- Anemia, weight loss, fatigue, rectal bleeding
- Increased thirst, increased urination, fatigue
- Infant born to mom w/ Sjogren's or Lupus
- Form of vasculitis, peeling hands/feet, high fever, red eyes, lymphadenopathy

Hematologic Autoimmune Diseases

- Autoimmune Hemolytic Anemia
- Henoch-Schonlein purpura (HSP)
- Autoimmune Neutropenia
- Immune thrombocytopenic purpura (ITP)

**Symptoms:**
- Pallor, fatigue, dark urine
- Acute abdominal pain, purpuric rash on buttocks/upper thighs/feet and ankle
- Recent viral infection, petechial rash, nosebleed

Goals of Treatment

- Reduce inflammation/relieve symptoms
- Prevent damage
- Replace vital substances no longer made
- Optimize quality of life
- Risk/benefit
Discovery of Dual Therapies

Mustard Gas in July 1919 recognized as immunosuppressive
Immunosuppressive properties for both cancer therapy and diseases with heightened/abnormal immune activity
Cytotoxic drugs inhibit the immune system as part of their action
Challenge is predicting the patients that will benefit from particular therapeutic approaches.
Combining biologic therapies with traditional chemotherapy drugs is more effective than either type of therapy alone

Non-Biologic Treatment Options

<table>
<thead>
<tr>
<th>NSAIDS</th>
<th>Immunomodulators</th>
<th>Common Adverse Effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Naprosyn, Ibuprofen</td>
<td>Mercaptopurine (6MP)</td>
<td>Nausea, vomiting, anorexia</td>
</tr>
<tr>
<td>Corticosteroids</td>
<td>Azathiopurine</td>
<td>Headache</td>
</tr>
<tr>
<td>Prednisone, Methylprednisolone</td>
<td>Methotrexate</td>
<td>Nausea, vomiting, anorexia</td>
</tr>
<tr>
<td>DMARDS, non-biologic immunosuppressants</td>
<td>Methotrexate</td>
<td>Rash, sun sensitivity</td>
</tr>
<tr>
<td>Sulfasalazine</td>
<td>Cyclophosphamide</td>
<td>Weight gain/loss</td>
</tr>
<tr>
<td>Hydroxychloroquine</td>
<td>Cyclosporine</td>
<td>Fatigue</td>
</tr>
<tr>
<td>Methotrexate</td>
<td>Tacrolimus</td>
<td>Fatigue, nausea, vomiting, anorexia</td>
</tr>
<tr>
<td>Sulfasalazine</td>
<td>Cyclophosphamide</td>
<td>Increased risk of infection</td>
</tr>
<tr>
<td>Chemotherapy</td>
<td>Cyclophosphamide</td>
<td>Hypersensitivity</td>
</tr>
<tr>
<td>Blood Products</td>
<td>Cyclophosphamide</td>
<td>Renal and/or hepatic toxicities</td>
</tr>
</tbody>
</table>

Common Adverse Effects:
- Nausea, vomiting, anorexia
- Headache
- Rash, sun sensitivity
- Weight gain/loss
- Fatigue
- Increased risk of infection
- Hypersensitivity
How Biologic Therapy works

Targets
- Immune system response
- Injectable
- Few side effects

Biologic DMARDS

- Etanercept (Enbrel)
- Infliximab (Remicade)
- Adalimumab (Humira)
- Rituximab (Rituxan)
- Anakinra (Kineret)

There are little or no standards in place
- Increased risk of infection
  - Hold medication if signs of infection

Consider these patients immunocompromised

Injection site and/or infusion reactions
- Apply supportive measures, do not hold

Little data for pre-surgery
- Usually not necessary to hold for dental
- No live vaccines during therapy

Initiating Biologic Therapy in Autoimmune Diseases

- Complete set of lab work
  - Tuberculosis screening
  - Hepatitis B/C screening
  - Liver Function Tests
  - CBC
  - Lipo (for tocilizumab, infliximab)
  - Urine
- Vaccinate per CDC, live vaccines within 4 weeks
- Pharmacists education and injection training
- Discuss Pregnancy and Breastfeeding
- Geographics
There are currently few or no standards in place. Signs and symptoms of infection require hold therapy for infections. Injection-site and/or infusion reactions may occur. Consider repeating PPD and Hepatitis screening and for elective surgery. No data on when to hold therapy. Usual practice: stop 1 to 1.5 doses ahead of time (e.g., stop etanercept 1 to 1.5 weeks ahead, adalimumab 2 weeks ahead). Usually not necessary to stop therapy for dental procedures or colonoscopies.

**Consequences of stopping therapy**
- Return of inflammation
- Permanent joint damage
- Organ damage/failure
- Disfigurement
- Increased health care costs

**Differences in Monitoring Practices**

<table>
<thead>
<tr>
<th><strong>Oncology</strong></th>
<th><strong>Autoimmune Disease</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Limited to duration of therapy</td>
<td>Lifelong</td>
</tr>
<tr>
<td>Lab frequency r/t chemo-pancytopenia</td>
<td>Labs every month-3 months</td>
</tr>
<tr>
<td>Frequent blood component transfusions</td>
<td>Self administration/monitoring</td>
</tr>
<tr>
<td></td>
<td>Rare blood component transfusions</td>
</tr>
<tr>
<td></td>
<td><strong>Can become neutropenic</strong></td>
</tr>
</tbody>
</table>

**Return of inflammation**
- Increased health care costs
School Considerations

- School provides structure, normalcy, sense of purpose.
- Adults have jobs in order to live, children have school.
- 504 Plans:
  - Access to discrete bathrooms.
  - Tutoring for missed classes due to prolonged absence.
  - Extra time to make up for missed testing/assignments.
  - Access to adequate hydration.
  - Use of laptops for class notes.
  - Extra set of textbooks for home.

Challenges faced by these patients:
- Changes in physical appearance (steroid therapy).
- Weight gain or loss.
- Altered self-image.
- Anxiety, social isolation, depression.
- CID.
- Inability to fully participate in activities with their peers.
- School absences due to clinic visits/infusion therapy.

Safety Considerations

Patients on biologic therapies should avoid:
- Fish aquariums or handling reptiles.
- Cleaning animal and bird cages (bird droppings).
- Pet/animal scratches.

Handwashing after recess/playgrounds:
- Gloves if working in soils (gardening).
- Gloves when working with pond or stagnant water activities (science activities).

Any breaks in skin caused by pet/animal scratches should be cleaned and monitored for signs of infection.
If someone is on a biologic, and has signs of an infection or is seen in ED:
  - Assume the patient is immunocompromised and is at risk for infection
and is sent home from ED or discharged:
  - Make sure the patient knows to contact his/her Rheumatologist or Specialty Pharmacist about whether to resume usual dosing schedule for biologic, especially if the patient is being sent home on antibiotics!

Take Home For Patients on Biologic Therapy

If there is an outbreak of a communicable disease, please remember to notify these parents so they can seek medical advice.
- Children on biologics should not receive live vaccines
- All other vaccines should be up to date, annual flu immunization in the fall (no nasal mist)

Take home cont’d

Reality Bites
The Future

Challenges in predicting which patients could benefit from particular therapies
DNA technology to identify specific genes
Relevance in commonalities in pathogenesis
Use of more targeted therapies earlier in the disease
Development of regimens to induce remission

Special Thanks

Amanda Kennedy  
Clinical Pharmacist  
Rheumatology

Jill Sullivan, MD  
Pedi GI

Leslie Abramson, MD  
Pedi Rheumatology

References

References:  
American Autoimmune Related Diseases Association  
www.aarda.org/
Questions?