

IDF Lunch & Learn: Good's Syndrome

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Today's Topics

What is Good's Syndrome?

1. What is a **thymus** and a **thymoma**?
2. What are **B cells** and **antibodies**?
3. What are **T cells**?

What to watch out for in Good's Syndrome and thymoma?

Living with Good's Syndrome

1. What are the **treatments**?
2. Navigating the **pandemic** with immunodeficiency

Q and A

First report of Good's syndrome (1955)

Mr. F.H. is a 58-year-old male who was well but then developed pneumonia for the first time ... chest [x-ray] revealed evidence of a large thymoma.

On November 6, 1951, a 54 gram tumor was removed from the anterior mediastinum ... benign thymoma ...

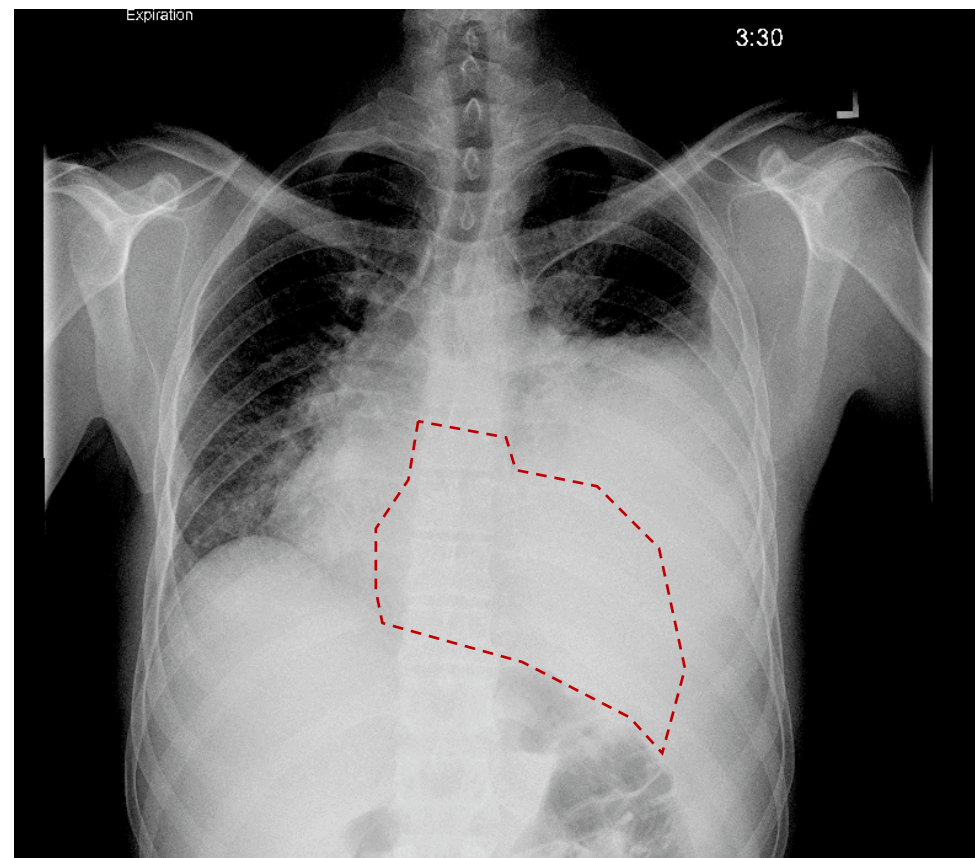
He then had 17 pneumonias in the next 4 years. He had an absence of [antibodies].



Figure 1 Robert A. Good (1922–2003).

A case from Mount Sinai Hospital

- 51 year old architect
- Long history of sinusitis, but has not required hospitalization
- Felt poorly and fainted. His family doctor thought his chest sounds were dull
- Referred for chest X-ray - thymoma was found and surgically removed
- 8 months later, he went to an allergist for ongoing sinusitis
- Allergist tested antibody levels:
IgG 30, IgA 0, IgM 5 mg/dL
- Patient referred to Mount Sinai Immunology



Thymoma

Antibodies
B cells

Good's Syndrome

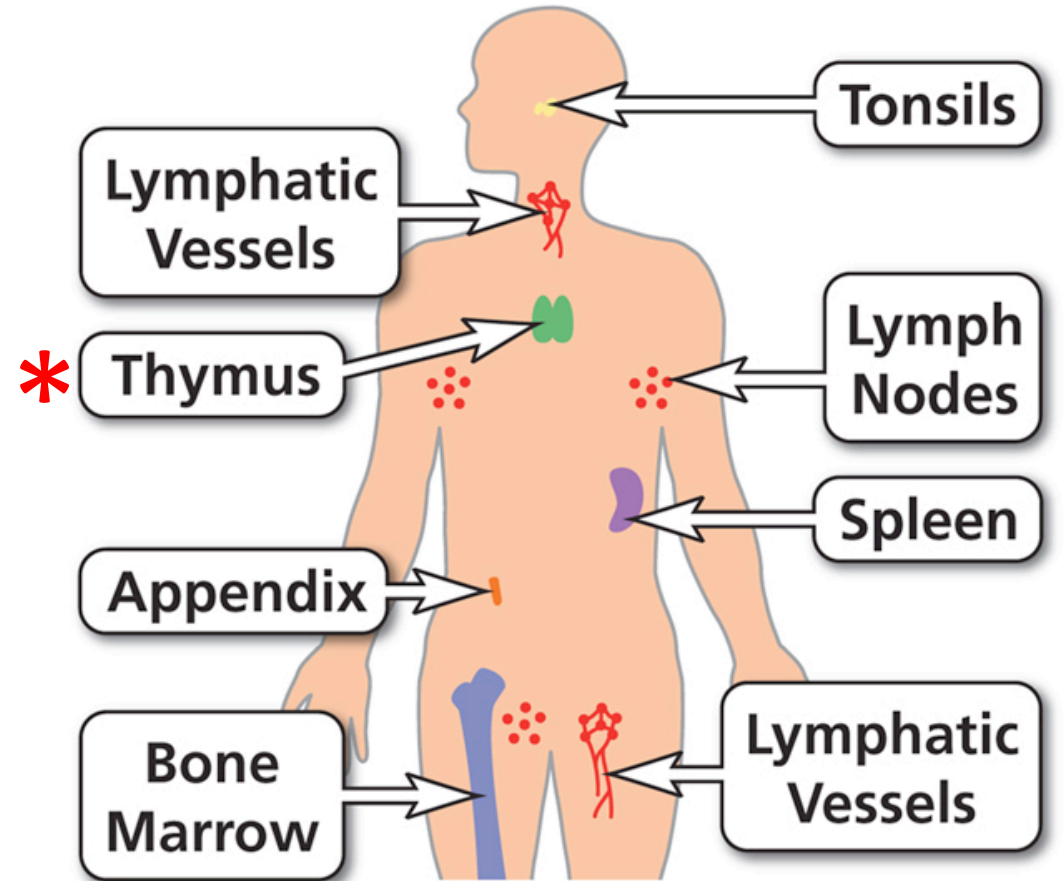
T cells

Autoimmunity

What is a **thymus**?

- A gland below the chest
- Large in infants, but small in adults
- “Thumos” (thymus) is a Greek word for “spiritedness” or seat of the soul

Immune System



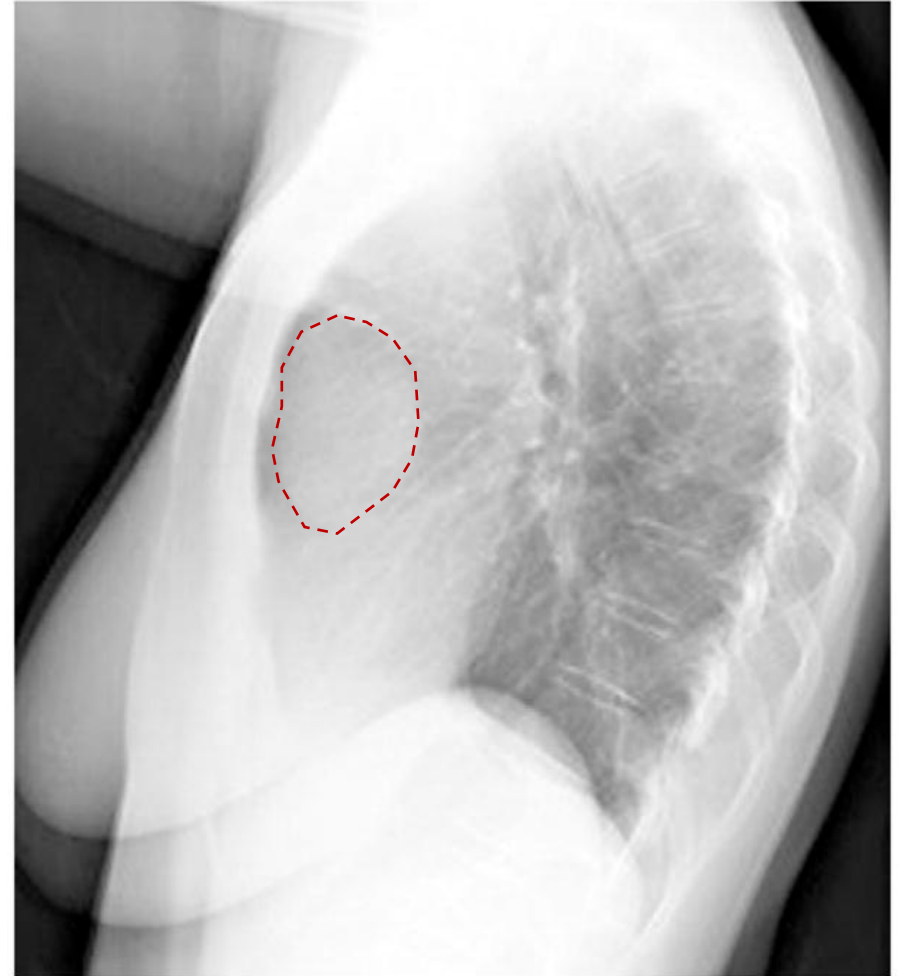
Thymus: a school for immune cells (T cells)

- In the thymus, T cells learn to attack microbes (bacteria, viruses, fungi).
- Bad T cells that can attack our own cells are also removed.
- The immune system produce most of its T cells during childhood.



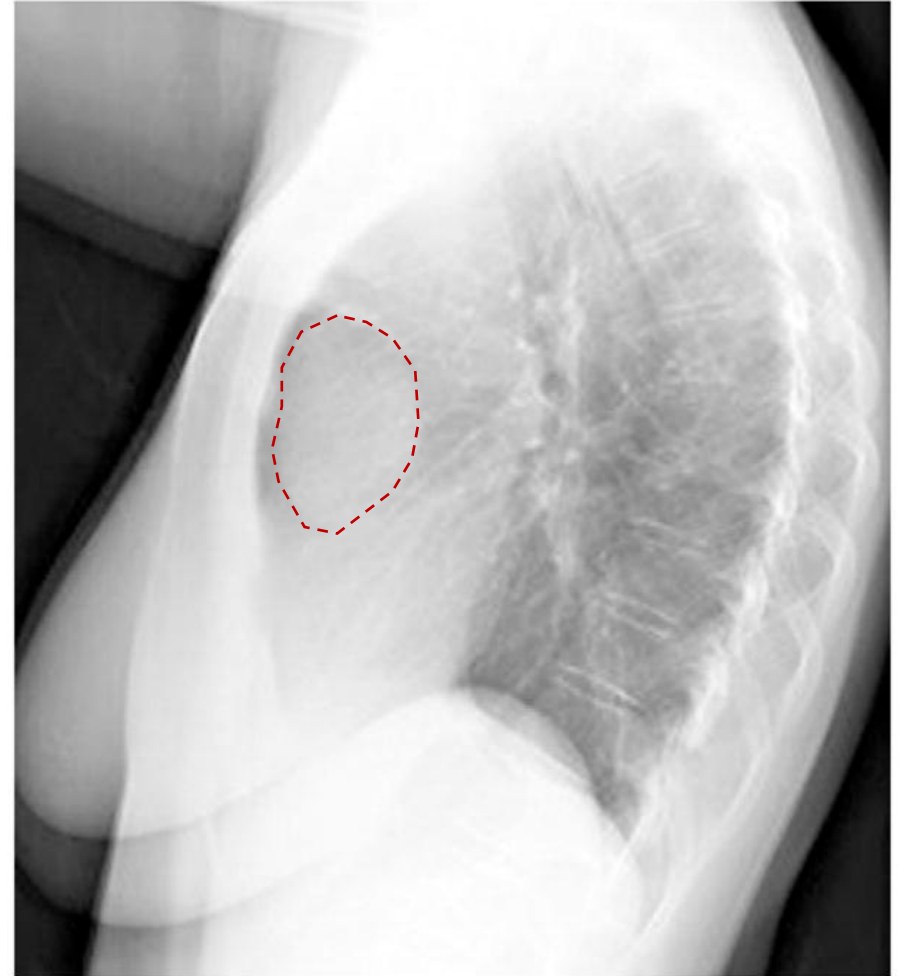
What is a **thymoma**?

- A rare tumor of thymus gland
- Typically occurs in 4th-6th decades of life
- No known risk factors
- Slow growing
- Often benign (sometimes malignant)



What are symptoms of **thymoma**?

- Can be an incidental finding only
- Tumor compresses on heart and lungs (chest pain, cough, shortness of breath)
- An associated syndrome:
 - Myasthenia gravis (muscular weakness)
 - Red cell aplasia (anemia)
 - **Immune deficiency: $\sim < 5\%$ low antibodies (Good's syndrome)**



Our Immune System



Thymoma

Antibodies
B cells

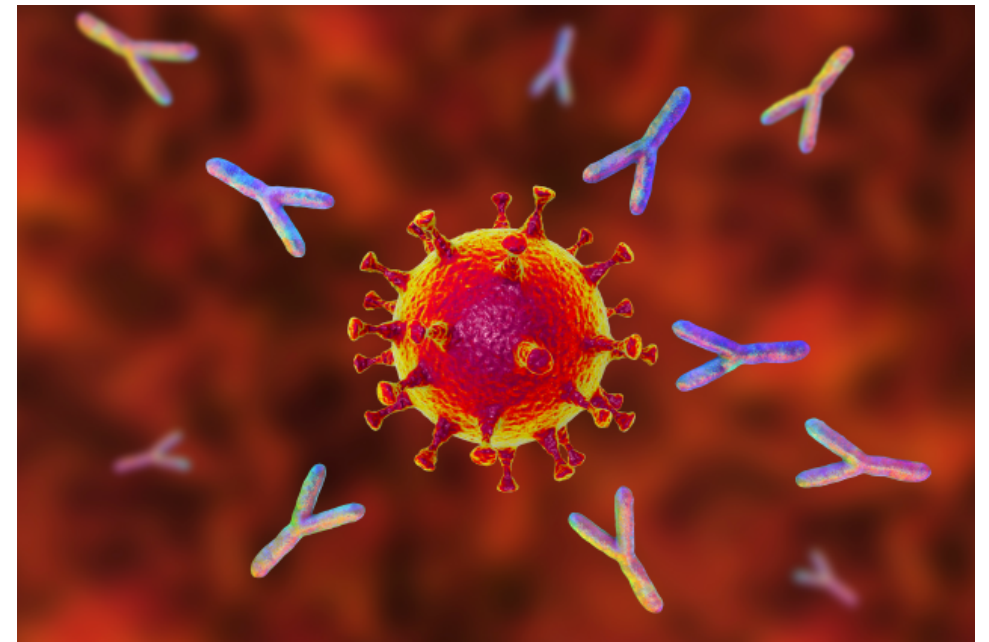
Good's Syndrome

T cells

Autoimmunity

Antibodies: Missile Defense

- Antibodies protect us when unwanted substances enter our body
- Antibodies fight against: bacteria, viruses, fungi
- Produced by specialized immune cells called **B cells**
- In Good's Syndrome: B cells are very low or absent.



Thymoma

Antibodies
B cells

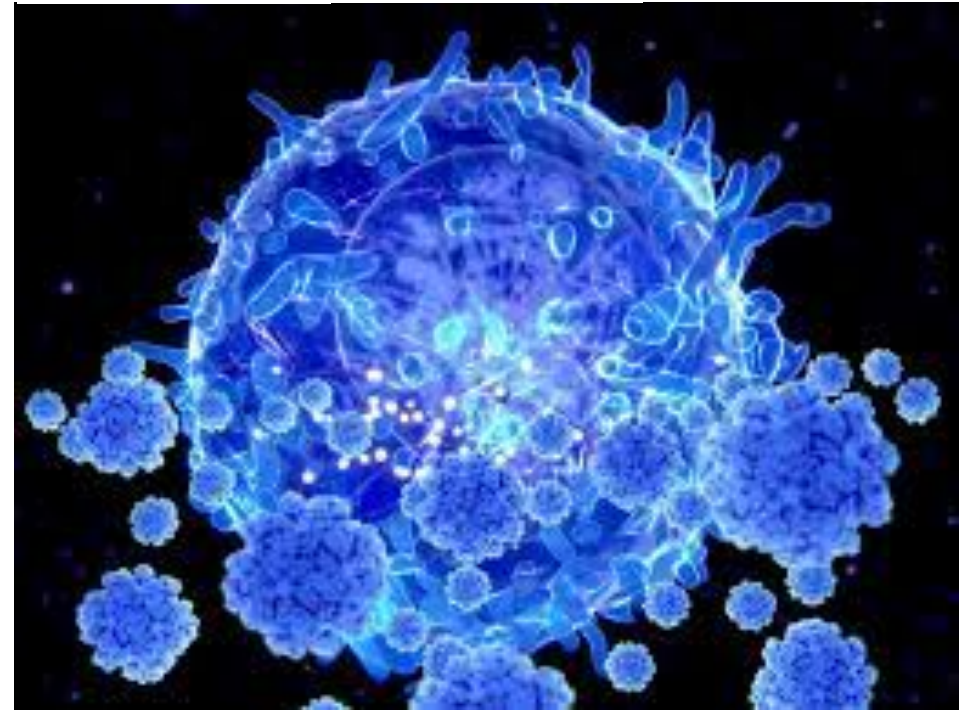
Good's Syndrome

T cells

Autoimmunity

T cells: Immune System's Quarterback

- Trained at thymus
- Orchestrate targeted immune responses
- Especially important for viruses, fungi



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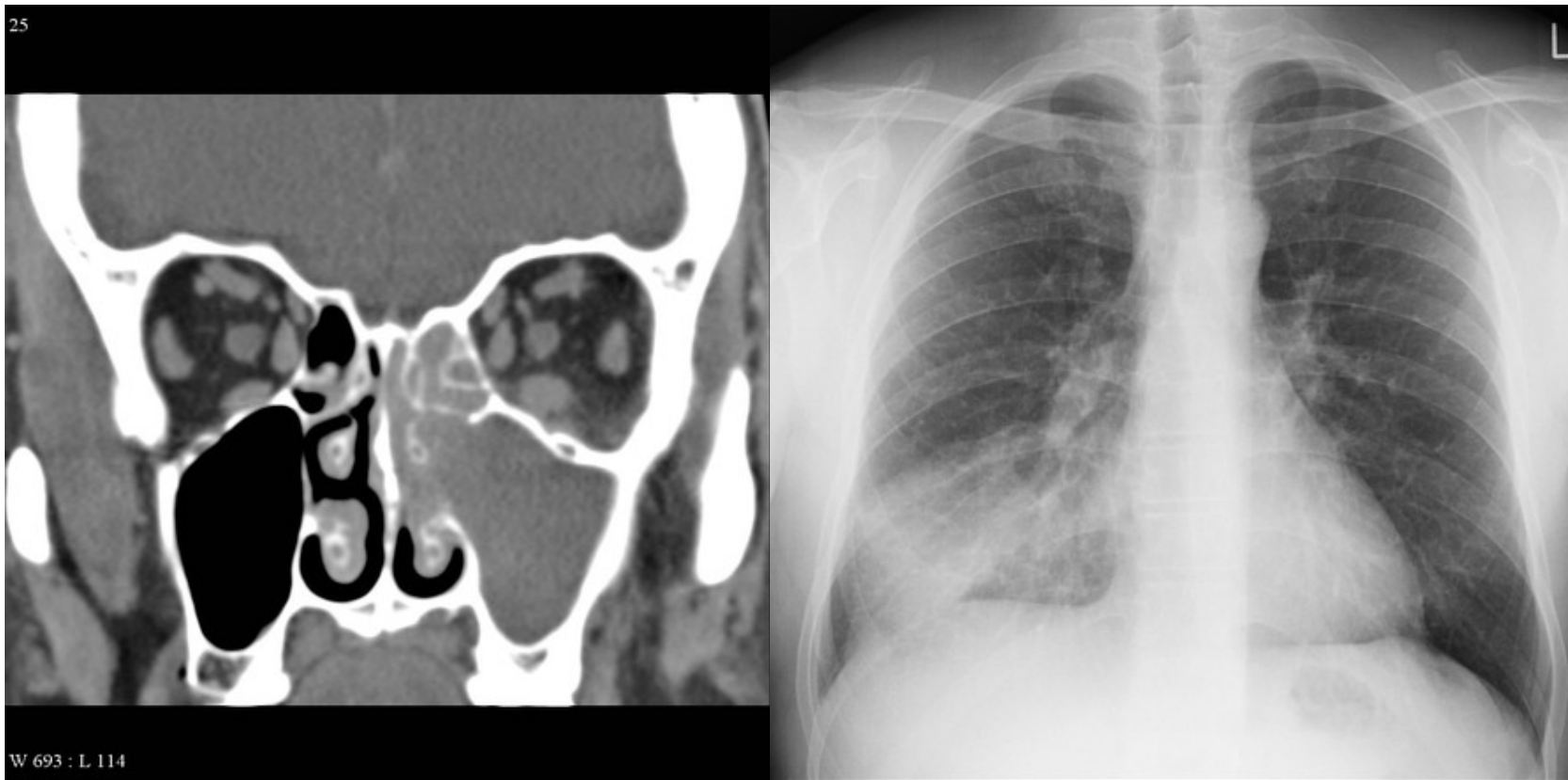
What to watch out for in Good's Syndrome and thymoma?

Living with Good's Syndrome

1. What are the treatments?
2. Navigating the pandemic with immunodeficiency

Q and A

1. Infections



UK report (78 patients)	
Feature	%
Bronchiectasis	45%
Sinusitis	10%

1. Infections

Bacteria	Viruses
Haemophilus	Cytomegalovirus
Streptococcus	Herpes simplex
Staphylococcus	Varicella zoster
Salmonella	Human herpesvirus 8
Campylobacter	
Mycobacteria	
Fungi	Protozoa
Candida	Giardia
Pneumocystis carinii	

Thymoma

Antibodies
B cells

Good's Syndrome

T cells

Autoimmunity

2. Autoimmunity

UK report (78 patients)	
Feature	%
Autoimmunity	26%
Low red blood cells	10%
Thyroid disease	8%
Arthritis (joint inflammation)	5%
Myasthenia gravis	4%
Lupus	4%
Sjogren's disease	3%

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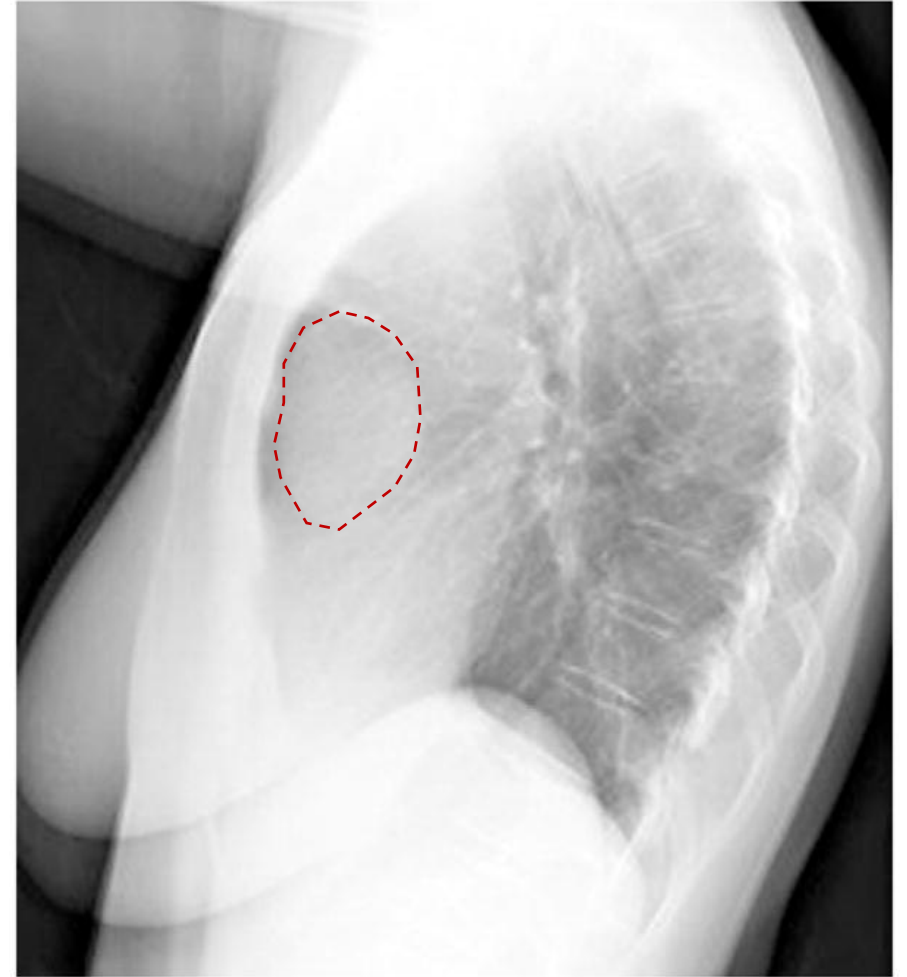
Living with Good's Syndrome

1. What are the treatments?
2. Navigating the pandemic with immunodeficiency

Q and A

Good's Syndrome: Treatments

1. Remove thymoma
 - Rarely cancerous, but possible
 - Removal of thymoma does not restore the immune system



Good's Syndrome: Treatments

1. Remove thymoma
2. Replace antibodies
 - IVIG (monthly) – infusion center or home
 - SCIG (weekly, or monthly) - home



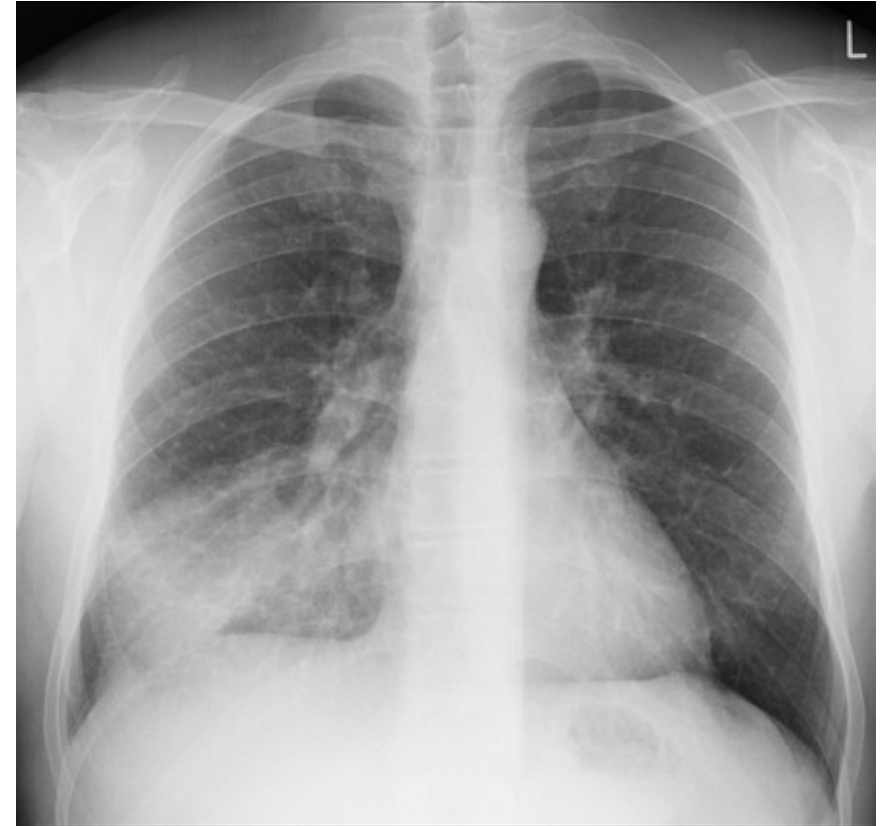
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Good's Syndrome: Treatments

1. Remove thymoma
2. Replace antibodies
3. Treat infections
 - Antibiotics
 - Antivirals
 - Consider prophylaxis if pre-existing lung disease



Good's Syndrome: Treatments

1. Remove thymoma
2. Replace antibodies
3. Treat infections
4. If autoimmunity
 - Work with specialists for targeted treatments



Good's Syndrome Treatment Tips

1. Check in with your immunologist routinely

- Tell them about any interval infections, weight changes, new symptoms, and new medications
- Check your antibody levels 1-2 times a year

2. Protect yourself from infections

- Optimize IVIG/SCIG
- Test broadly if symptomatic: bacteria, viruses, fungi, etc
- Protect your lungs
- If sinusitis, work with an ENT physician to consider topical treatments

3. If you develop new symptoms, tell both your primary care doctor and immunologist

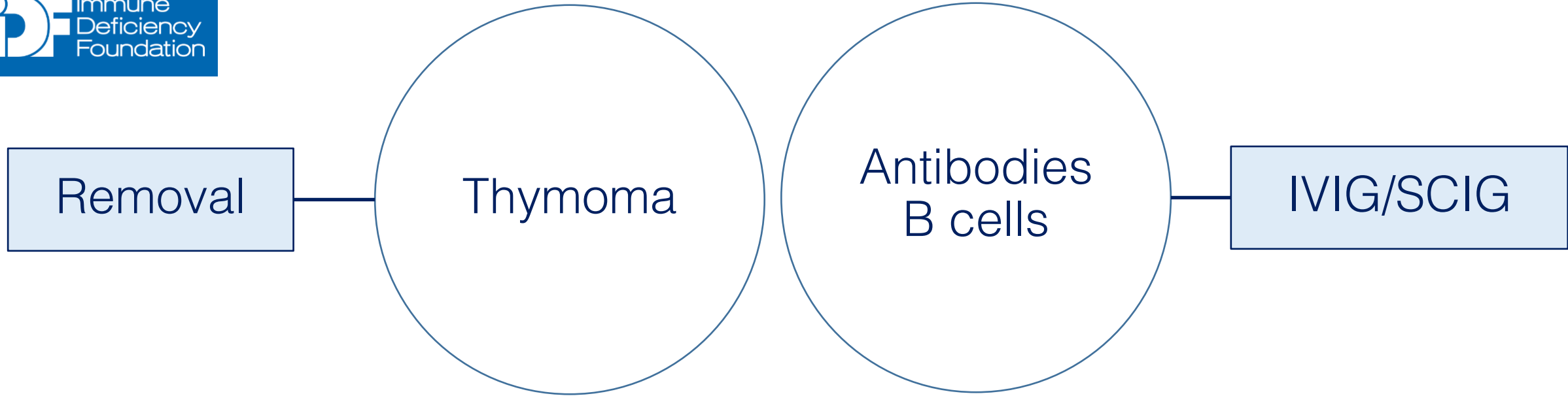
- Thymoma can be associated with autoimmune diseases

Covid – general advice

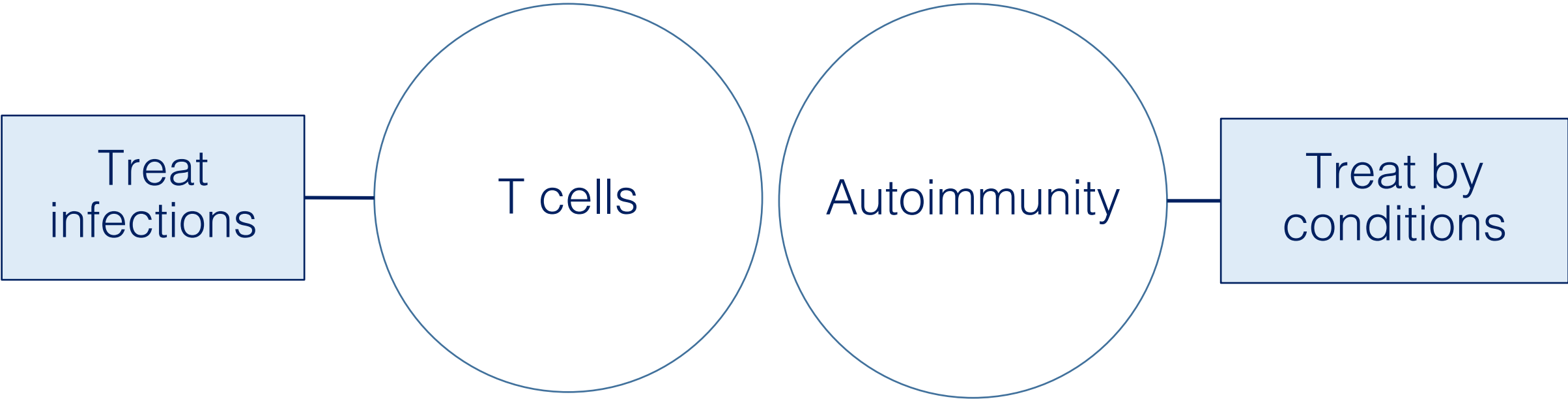
- Vaccinations and masks
- Antiviral therapies can prevent progression to severe disease
 - Paxlovid® is currently approved under EUA
 - Works well if given within 5 days of symptoms onset
 - Ask your doctor and pharmacist to check for drug interactions
- The efficacy of monoclonal antibodies varies depending on the dominant covid variant
 - Check in with your immunologist on the most up-to-date information

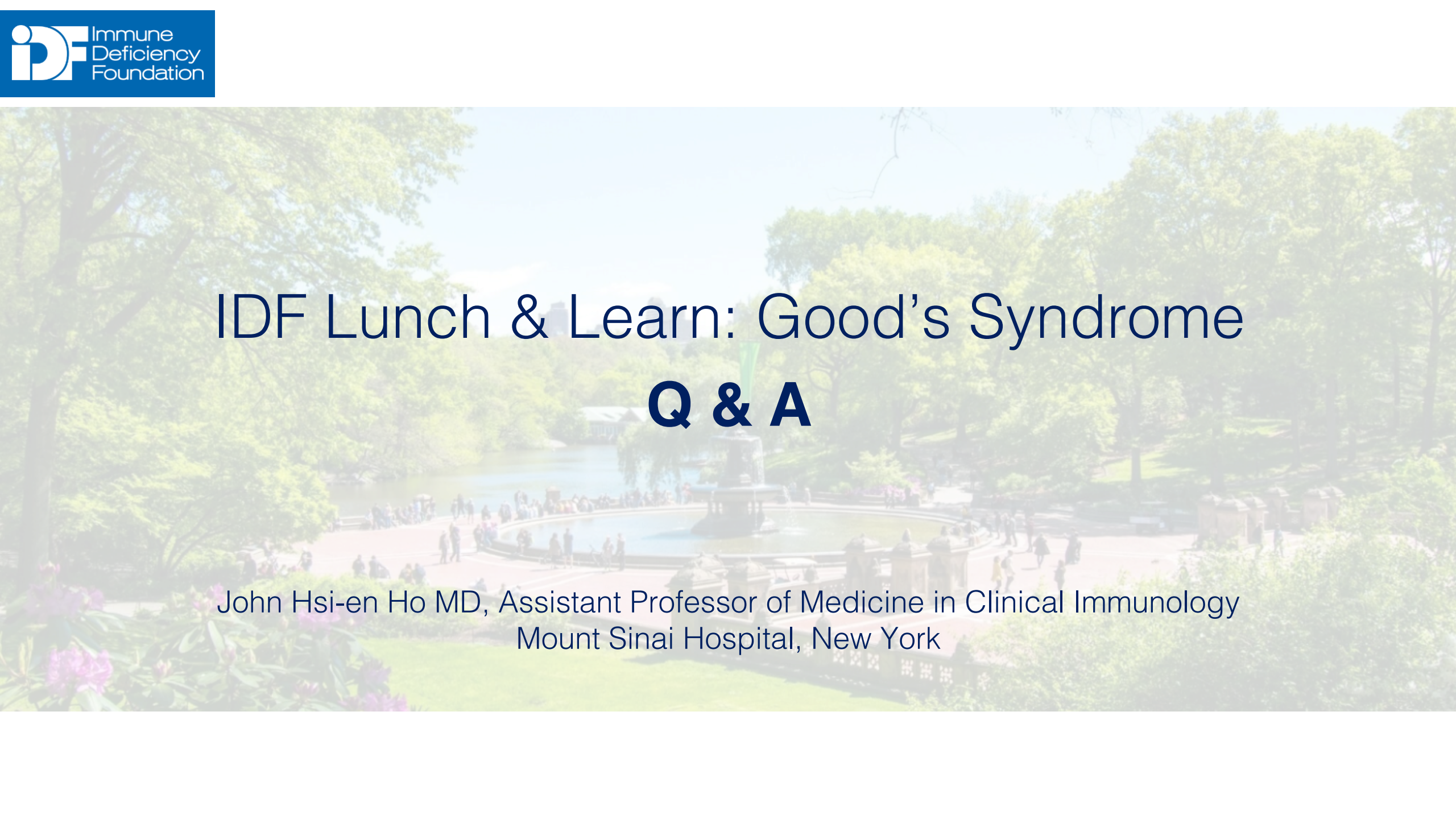
Good's Syndrome: Summary

- Good's syndrome is a rare, acquired immune deficiency
- Characterized by previous or current thymoma
- Low antibodies and B cells are characteristic. Removal of thymoma does not restore this.
- T cells can also be affected
- Symptoms include infections and autoimmunity – causes of the immune defects are still under investigation



Good's Syndrome



The background of the slide is a photograph of Central Park in New York City. It shows a large, circular fountain with water spraying upwards, surrounded by a paved walkway where many people are walking. The park is filled with lush green trees and grass, and a small body of water is visible in the background under a clear blue sky.

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Q & A

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