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Horse or Zebra? The Common and Uncommon Presentations of CVID

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Horse or Zebra?
The Common and Uncommon Presentations of CVID

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Introduction

•Common Variable Immunodeficiency (CVID) is the most frequently encountered primary immunodeficiency. It consists of a group of heterogenous antibody deficiencies which most commonly predisposes patients to recurrent respiratory infections. However, some patients do present with other features of a weakened immune system such as autoimmune. diseases and/or malignancies.[1]
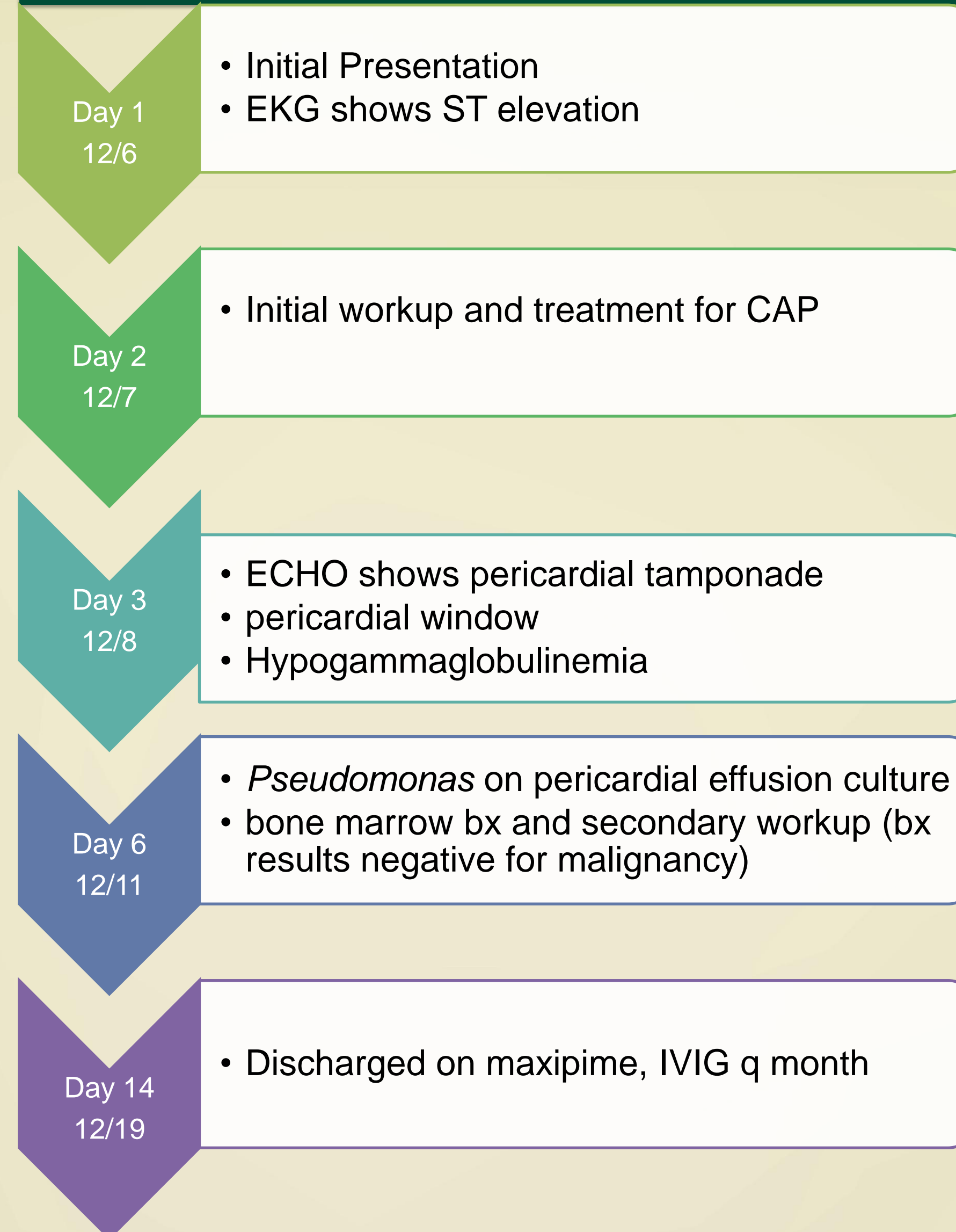•Disease of B cell differentiation failure.

Patient Information

1. 47 year old Hispanic male presents to the ED with three-day history of dyspnea, fever, chills and cough productive of clear sputum.
2. No significant past medical history until the preceding year, when he was hospitalized 3-4 times with a diagnosis of pneumonia.
3. He works as a truck driver traveling in the southwest United States, but primarily in Arizona.
4. The patient reported temporary resolution of symptoms with each antibiotic treatment (name unknown).
5. He also reports persistent conjunctivitis for the past year.
6. He denies any significant childhood illness or hospitalization.
7. He admits to two casual sexual contacts in the last year.

Admission

•Temp 98.5
•BP 74/46
•HR 118
•Dyspnea with extended conversation; diffuse coarse rhonchi with diminished air exchange in bilateral bases
•Conjunctivae mildly injected with purulent matted discharge in the upper eyelashes
•WBC 35,500 with predominance of neutrophils

Timeline

Day 1 12/6
• Initial Presentation
• EKG shows ST elevation

Day 2 12/7
• Initial workup and treatment for CAP

Day 3 12/8
• ECHO shows pericardial tamponade
• pericardial window
• Hypogammaglobulinemia

Day 6 12/11
• *Pseudomonas* on pericardial effusion culture
• bone marrow bx and secondary workup (bx results negative for malignancy)

Day 14 12/19
• Discharged on maxipime, IVIG q month

Diagnostic Assessment

•Serology: complement levels, Immunoglobulins, rheumatoid screen, Coccidiomycosis antibody, Histoplasma antibody, HIV screen negative, quantiferon negative
•Bone marrow biopsy negative
•EKG: diffuse ST elevations
•ECHO: pericardial tamponade
•Pericardial fluid culture: *Pseudomonas*
•Sputum culture: respiratory flora
•Urine culture: negative
•Conjunctival fluid culture: normal flora
•CXR: opacities at lung bases
•Chest CT: bibasilar pneumonias, extensive mediastinal adenopathy, pericardial effusion
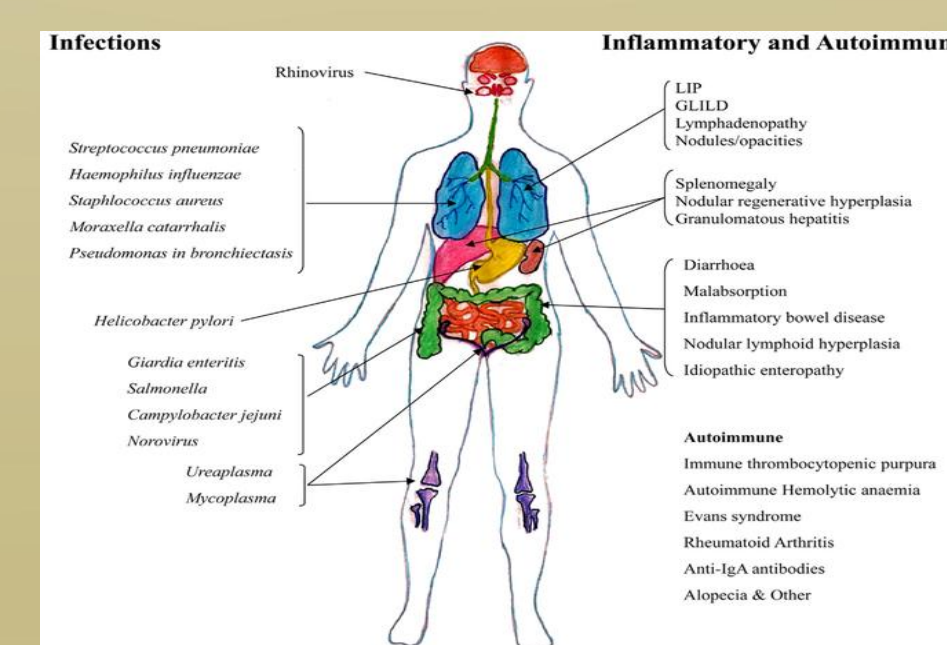
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Therapeutic Interventions

•Pericardial window performed to alleviate pericardial tamponade.
•IVIG infusions initiated during hospitalization.
•Azactam given, then changed to Zosyn for *Pseudomonas* coverage.
•Maxipime on discharge, with plan for monthly IVIG infusions.

Follow Up Outcomes

•Summarize clinical course of all follow up visits including
  -Clinician and patient assessed outcomes
  -Intervention adherence and tolerability
  -Adverse and unanticipated events

Discussion

•This case demonstrates the wide range of presentations with CVID. Quick diagnosis is imperative to avoid delays in treatment.
Incidence
•1:10,000 - 1:50,000 [2]
•Equally affects both genders
•34% of patients were diagnosed before the age of 10
•Typical delay in diagnosis of 4-9 years [3]
Common Presentations
•CVID is the most common primary immunodeficiency.
•It most commonly presents during puberty and young adulthood.
•Although the presenting symptoms vary widely, it most commonly manifests itself as recurrent, bacterial, upper or lower respiratory tract infections.
•Other presentations include conjunctivitis, GI infections, chronic lung disease, autoimmune disease, GI inflammatory disease, and malignancies
•By definition, CVID causes low to absent levels of immunoglobulins, specifically IgG, IgA and/or IgM, in the absence of any other immunodeficiency state. As such, it is associated with poor response to immunizations. [4]
Treatments
•CVID is primarily treated with IVIG which has been shown to decrease pulmonary infections and hospitalizations. [5]
•IVIG is given at a dose of 300 to 400mg/kg every 3-4 weeks. Trough IgG levels are measured every 6 months after the first dose. [5,6]
•Higher doses can be given if patients have continued major infections or chronic lung disease. [6]
•Patients on IVIG tend to remain susceptible to sinusitis and gastrointestinal infections. [7]
•Infections should be treated with antibiotics. Other associated conditions (e.g. autoimmune disorders, enteropathies, granulomatous disorders) can be treated symptomatically.
•Patients should receive age appropriate cancer screenings and must be monitored for malignancies including lymphoma and gastric cancer. [8,9,10]
•Radiation exposure should be limited due to the elevated risk of developing malignancy
•With the use of IVIG, the major causes of death are complications of chronic lung disease and malignancies, which can shorten the life span significantly (mean age: mid-50's). [11,12]

KEEP CALM AND IVIG ON