

**DIFFERENT THERAPEUTIC  
APPROACHES IN PATIENTS  
WITH CHRONIC  
GRANULOMATOUS DISEASE**

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# Infectious deteriorations in CGD patients

25 CGD patients observed over 13 years

- ✓ Staphylococcus aureus infections (liver abscess, destructive pneumonia, lymphadenitis, osteomyelitis)
- ✓ Mycobacterial infections (BCG-osis, lymphadenitis, tuberculosis, disseminated mycobacteriosis, osteomyelitis);
- ✓ Aspergillosis (pulmonary, brain involvement)

# **THERAPY**

## **in most frequent cases**

- ✓ Preventive antimicrobial therapy:
  - ✓ Co-trimoxazole 5mg/kg (for trimetoprim);
  - ✓ Itrakonazole 5-8mg/kg.
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- ✓ Infectious deterioration's therapy:
  - ✓ Antibacterials (wide spectrum with intracellular penetration) – long term
  - ✓ Antimycotic (Amphotericin B, Fungizone, Vorikonazole) - long term
  - ✓ Antituberculosis (3, 4 medications) – long term

# **THERAPY**

## **in severe cases**

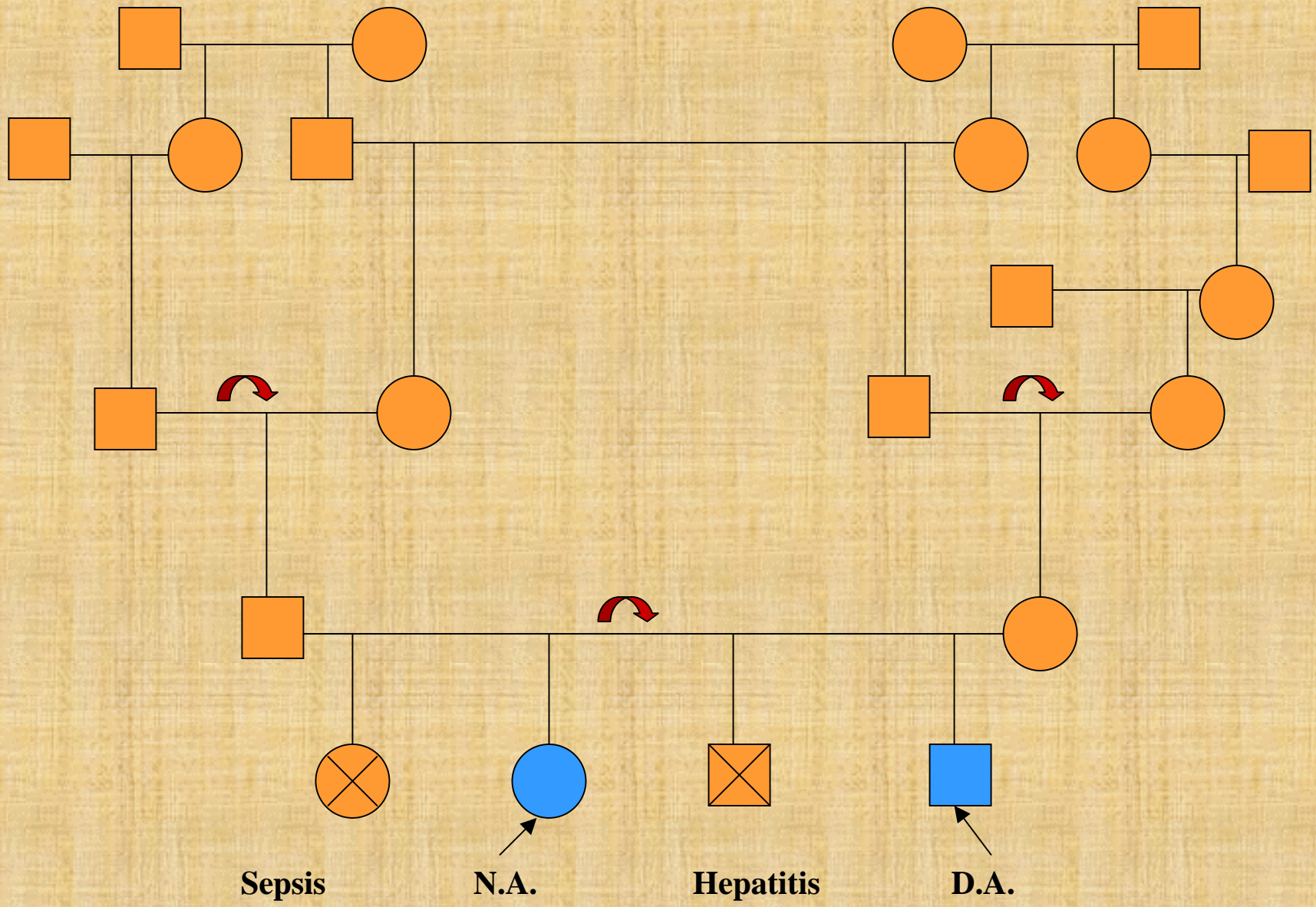
- ✓ Granulocyte stimulating factor (GCSF) – 3-5mkg/kg  
3 times a week – long term;
- ✓ Granulocytes transfusions – 3 times a week – 2-3  
weeks;
- ✓ Recombinant human Interferon-gamma – 50mkg/m<sup>2</sup>  
3 times a week;
- ✓ Surgical treatment (for refractory infectious sites).

# **N.A. – 11 y.o Chronic Granulomatous Disease**

## **Disseminated mycobacterial infection, mycobacterial C2 and occipital osteomyelitis**

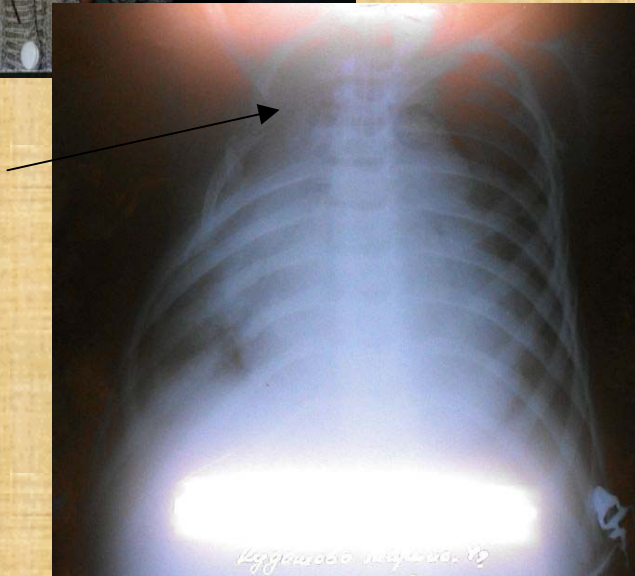


- ✓ 4 mo - BCG-osis, cervical lymphadenitis;
- ✓ 7 mo - fistulas formation
- ✓ 12 mo - CGD diagnosis;
- ✓ 2 y.o. - Disseminated BCG-osis;
- ✓ 9 y.o. -deterioration of mycobacteriosis – osteomyelitis, spondilitis, open fistulas;
- ✓ PCR Mbt tuberculosis positive;
- ✓ Admission of specific qudritherapy for years + GCSF+Recombinant IFNg with partial clinical effect.



# **M.K. 4 y.o. – Chronic Granulomatous Disease**

## **Disseminated mycobacterial infection, mycobacterial osteomyelitis, paravertebral abscesses**

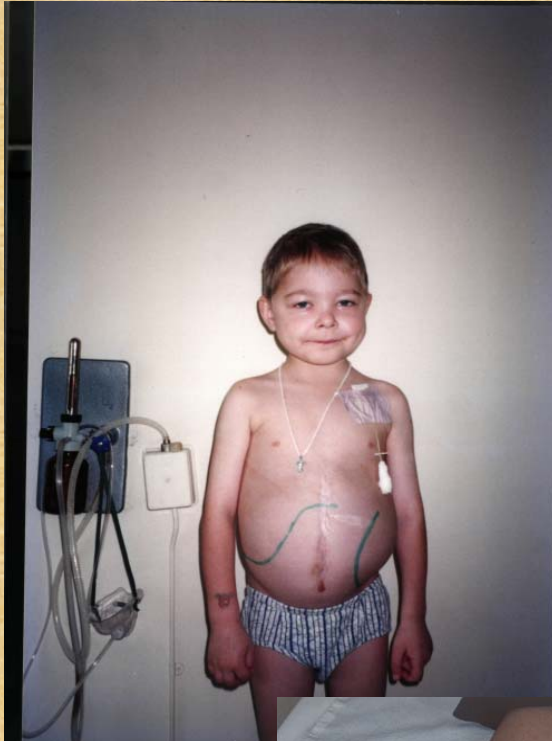


- ✓ 2 y – Multifocal tuberculosis of lymph nodes, lungs, pleura, peritoneum, liver, spleen. CGD diagnosis suspected;
- ✓ 4 y - evaluation of the above diagnoses;
- ✓ Deterioration of mycobacteriosis – osteomyelitis, spondilitis;
- ✓ PCR Mbt tuberculosis positive;
- ✓ Admission of wide spectrum antibiotics, antimycotics, specific pentatherapy for 4 months + granulocytes transfusion showed partial clinical effect.
- ✓ Surgical approach - toracoplastic - no effect.

# **E.K. - 6 y.o. Chronic Granulomatous Disease**

## **Chronic X-XII ribs osteomyelitis**

### **Aspergilous arthritis**



- ✓ **6 mo - BCG-osis;**
- ✓ **2 y.o. - hepatosplenomegaly;**
- ✓ **3 y.o. - multiple liver abscesses;**
- ✓ **6 y.o. - soft tissue abscess in paravertebral area of mixed bacterial/mycobacterial origin deteriorated by ribs osteomyelitis;**
- ✓ **Intensive antibacterial and antifungal treatment for 3 months showed no effect but spread of infection;**
- ✓ **Surgical treatment along with granulocyte's transfusion provided excellent clinical effect.**
- ✓ **9 y.o. – prepatellar abscess of Aspergilla origin - intensive antibacterial, antifungal treatment +GCSF+surgical treatment - excellent clinical effect.**



**M.S. – 17 y.o. Chronic Granulomatous Disease**  
**Purulent mediastinitis, purulent epiduritis C7-D5.**  
**Multifocal aspergillosis**



- ✓ 2 wks – pneumonia;
- ✓ 2 mo – BCG-osis;
- ✓ 3 y.o. – lymphadenitis;
- ✓ 4 y.o – subdiaphragmal abscess;
- ✓ 6-8 y.o. – lymphadeitis relapse;
- ✓ 12 y.o. – pneumonia;
- ✓ 17 years – purulent mediastinitis, multifocal aspergillosis
- ✓ Intensive antibacterial and antifungal treatment for 5 months +granulocyte's transfusions+surgical approach provided good clinical effect