



Primary Immune Deficits in children: follow-up and outcome



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Background

Primary Immune Deficiencies (PID): rare under-determined diseases

Developed nations: 1/5,000 births/ year

Africa: 107,730 births/year (Bousfiha, Moroc 2012)

Purpose

Share following and outcome issue in PID patients

Methods

Type: Descriptive and analytical cross-sectional study

Site: Albert Royer Children's Hospital: 2014 - 2021

Conditions: HIV ruled out
Clinical diagnosis criteria

Biological examinations

Results

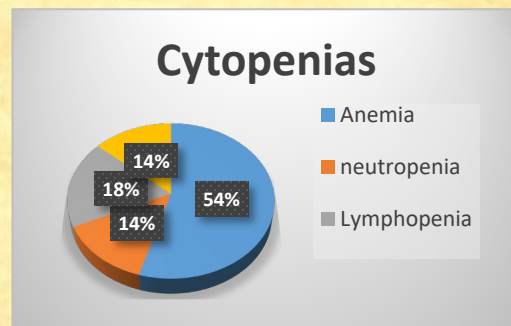
Included: 16/32

Sex-ratio: 0,6

Mean Dc age: 51,1months

Consanguinity: 50% (8/16)

Clinical signs: infectious, neurological, dermatological
Biological signs: cytopenias



Others signs:

hypogammaglobulinemia

Coombs Test

Bactériological exams



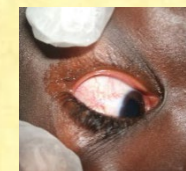
Results

Ataxie-Télangiectasie: 3/16

Lymphopenia:

TCD3 and BCD19

↑↑ αFP : 1161 ng/L



Verruciform

epidermodysplasia : 2/16

PNN= 540/ mm³

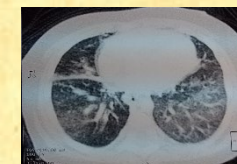
Typical histological aspect



Wiskott-Aldrich Sd: 1/16

SCID: 3/16

Type: T_ B_ NK+



Evans Sd : 1/16

Congenital neutropenia: 3/16

Hypogamma-globulinemia: 2/16

Chronic granulomatous disease: 1/16

Results

Outcome

- Remission flare: 6/16
- Bronchiectasis: 2/16
- Overall mortality: 37,5%
- SCID and ataxia mortality: 100%

Conclusion

PID: new approach of infectious diseases

Syndromic PID are more common in our patients

Reserved prognosis: SCID and ataxia



African Society for Immunodeficiencies
Société Africaine des Déficits Immunitaires
الجمعية الإفريقية لأمراض ضعف المناعة الأولى

PID-IEI IN AFRICA:
WHERE AND HOW TO FIND THEM?

Online 7th ASID Congress
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African School
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