長庚紀念醫院
Chang Gung Memorial Hospital

Immunodeficiency Algorithm by Functional Assessment

1. Recurrent infections:
   - In 1 year - OM x 4; Sinusitis x 2, Pneumonia x 2; or any 2 deep-seated infections (osteo, CNS, etc.)
   - Recurrent deep skin or organ abscesses
   - Persistent thrush (mouth or other skin sites) after 1y/o
2. Poor response to antibiotics:
   - Need for IV or prolonged course (>2 mo) to clear infections
3. Infection with signature organisms - PCP, *Serratia, Aspergillus, Nocardia, Pseudomonas*
4. Family history of Primary Immunodeficiency
5. Classic clinical presentation:
   - Ex. DiGeorge or Ataxia Telangactasia
6. Failure to thrive

LABS

- CH50, AH50, MBL – tests complement cascade function first (L72-220; M32-254)

- CBC/Differential
- Neutrophil oxidative burst assay H2O2 production (M32-253)
- CD11b/CD18 (M32-264)
- Phagocytosis (M32-263)
- Chemotaxis (M32-265)

- CBC/Differential
- Quantitative IgM, IgG, IgA, & IgE
- IgG subclass (L72-206, 208, 210,212)
- Anti-tetanus titers
- Anti-pneumococcal titers pre- & 4 wks post immunization
- Anti-A & B titer (L72-855)
- Immunfixation electrophoresis (L72-204)

- CBC/Differential
- T & B cell subset analysis (L72-266; 267; 272)
- DTH reaction to *Candida* & *BCG* (M32-266)
- Cytotoxicity (M32-262)

DEFECT TYPE

Complement

- Recurrent disseminated Neisserial infections.
- Pyogenic bacterial infections.
- Angioedema of face, hands, feet, or GI tract.
- Autoimmune symptoms (Lupus).
- Hisory suggestive of autosomal dominant inheritance.

Phagocytic

- Soft tissue abscesses or lymphadenitis.
- Infection with catalase + organisms (*Staph aureus, Serratia, E. coli, Aspergillus*).
- Poor wound healing.
- Delayed separation of the umbilical cord.
- Chronic gingivitis and periodontal disease.
- Mucosal ulcerations.

B-cell

- Recurrent bacterial sino-pulmonary infections or sepsis, particularly with encapsulated organisms.
- Chronic or recurrent gastroenteritis (Giardia and Enterovirus common).
- Chronic enteroviral menigo-encephalitis.
- Arthritis
- Unexplained bronchiectasis.

T-cell

- *Pneumocystis carinii* pneumonia.
- Fungal infections.
- GVHD (rash, abnormal LFT's, and chronic diarrhea).
- Recurrent, severe, or unusual viral infections.
- Failure to thrive.

Combined

Modified by Wen-I Lee MD, PhD

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Immunodeficiency Algorithm by Functional Assessment

1.反覆感染
- 一年內－中耳炎 4 次，鼻竇炎 2 次，肺炎 2 次，或任 2 次深部感染(如骨髓、中樞神經)
- 反覆性深部皮膚或器官膿瘍
- 一歲後持續性鵝口瘡(口腔或皮膚)

2.對抗生素反應不佳
- 需靜脈或延長的療程(>2 月)才能清除感染

3.感染特定微生物－PCP、Serratia、Aspergillus、Nocardia、Pseudomonas

4.厚發性免疫缺陷之家族史

5.特定臨床表現，例如 DiGeorage 症或毛細血管擴張性運動失調

6.生長遲滯

實驗室

- 全溶血補體活性、AH50、甘露糖結合凝集素(MBL)測試、補體層疊功能(L72-220、M32-254)

- 血液常規及血球分類計數
- 中性球氧化烈測試及過氧化氫生素(M32-253)
- CD11/CD18(M32-264)
- 吞噬作用(M32-263)
- 趨化作用(M32-265)

- 血液常規及血球分類計數
- 定量免疫球蛋白 M、G、A、E
- 免疫球蛋白 Imno IgG 次類別(L72-206、208、210、212)
- 抗破傷風抗體效價
- 抗肺炎鏈球菌效價(接種疫苗前及 4 週後)
- 免疫固定電泳(L72-204)
- 抗 A、B 血型抗體效價

- 血液常規及血球分類計數
- T、B 細胞分組分析(L72-266、267、272)
- 對念珠菌、卡介苗之延遲型過敏反應(M32-266)
- 細胞毒殺(M32-262)

缺損類型

補體
- 反覆瀰漫性 Neisserial 感染
- 化膿性細菌感染
- 臉、手、腳、腸胃道血管性水腫
- 自體免疫病(如狼瘡)
- 體顯性遺傳方式

吞噬細胞
- 軟組織膿瘍及淋巴腺炎
- 具觸酶之微生物(Staph aureus、Serratia、E. coli、Aspergillus)
- 傷口癒合差
- 臍帶延遲分離
- 慢性齒齦炎及牙周病
- 黏膜潰瘍

B-細胞
- 反覆細菌性鼻竇肺感染、敗血菌，尤其是具莢膜之微生物
- 慢性反覆腸胃炎(梨形蟲或腸病毒常見)
- 慢性腸病毒腦膜腦炎
- 無法解釋的氣管擴張症

T-細胞
- 肺囊蟲肺炎
- 黴菌感染
- 移植物抗宿主病(紅疹、不正常肝功能、慢性腹瀉)
- 反覆嚴重、不尋常病毒感染
- 生長遲滯