2007/01(審)

- Maternal SLE has a high prevalence of fetal wastage
 - 1. Spontaneous abortion
 - 2. IUGR
 - 3. Preterm delivery
 - 4. Still birth
 - 5. perinatal death
- Infants of maternal SLE are at risk for the **neonatal lupus syndrome**
 - 1. Pathophysiology: transplacental transfer of maternal IgG antinuclear antibodies; 以 anti-Ro

(SSA)為主,少數為 anti-La (SSB)

2. Clinical Manifestation

Transient	Permanent
Rash	Congenital complete heart block (CCHB)
Erythematosus, scaly	
Photosensitive	
Annular or elliptical	
Face and scalp	
Cytopenias	
Hemolytic anemia	
Thrombocytopenia	
Leukopenia	
Hepatosplenomegaly	
Myocarditis/periarditis	
? pneumonitis	
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PS: 除了 CCHB 會永久存在以外, 其他症會隨 maternal antibody 消失而緩解 (within several months)

- 3. Congenital complete heart block
 - 最常見的心臟異常疾病
 - Pathophysiology: depostion of immunoglobulin in fetal cardiac tissue; 以 anti-Ro (SSA)為主,少

數為 anti-La (SSB)

- Presenting finding: fetal dysrhythmia, pericardial effusion or hydrops
- Heart block is usually permanent
- Usually, Cardiac pacemaking should be instituted
- 4. Cutaneous lesion
 - 常以 widespread macular rashes 來表現
 - 偶而呈現 butterfly rash and discoid lesions
 - Generally appear within the first few weeks of life and disappear spontaneously within 6 months

- 病人出院時,應告訴家屬觀察是否有上述皮膚表現發生
- 5. Hematologic manifestation
 - Anemia and thrombocytopenia: rare
 - Thrombocytopenia: resolve over several weeks and unless there are bleeding manifestations, does not require treatment
- 6. Liver disease
 - Hepatic involvement occurs in at least 15% of infants with NLE
 - Clinical expression: hepatomegaly (with or without splenomegaly), generally with a picture of cholestasis
- Management of infant of SLE mother
 - 1. 產前已知有 fetal bradycardia→ 轉健保床
 - Examinations: complete EKG (for heart block evaluation), heart echo (for heart contractility and associated anomaly evaluation), evaluate any effusion or ascites, check anti-Ro and anti-La
 - 合併 hydrops fetalis: treated as hydrops fetalis
 - Indication of cardiac pacing: (1) congestive heart failure (2) ventricular rate below 55/min in isolated CCHB and below 65/min with associated heart disease (3) prolong QTc (usually > 0.50 sec) (4) Stokes-Adams attacks (5) frequent ventricular ectopic beats and (6) alternating ventricular pacemakers
 - Isoproterenol:
 - $0.05 \sim 0.10 \, \text{ug/kg/min}$
 - 功能: chronotropic effect and vasodilatory effect
 - 僅暫時用在無法放置 cardiac pacing 的情況,例如 EVLBW prematurity
 - 2. 產前未發現有 fetal arrhythmia
 - Complete PE: skin lesion, pale looking, petechia, ecchymosis, size of liver and spleen, heart rhythm→ 如果都正常→不用轉健保床
 - 觀察是否有 skin lesion, anemia, bleeding disorder, cholestasis, HSM ,這些症狀可能於出生後幾周至半年才出現。
 - 一定要告知家屬,出院後仍需注意是否有上述症狀,即 skin rash, petechia, pale-looking, jaundice, abdominal mass (distension or fullness)