

113 年奇美醫院胸腔內科臨床病例討論會

日期：中華民國 113 年 07 月 30 日(星期二)

時間及地點：16:00-17:00

課程活動題目：胸腔影像綜合討論會：Poland syndrome

主講人：蔣士仁主任

主辦單位：奇美醫院胸腔內科

課程地點：10 樓討論室

教育積分：台灣胸腔暨重症加護醫學會

參加對象：主辦單位所屬院內醫師

聯絡人：楊穎潔 (06-2812811-57132)

摘要：

Introduction:

Named after Sir Alfred Poland, Poland syndrome includes the features of ipsilateral breast and nipple hypoplasia and/or aplasia, deficiency of subcutaneous fat and axillary hair, absence of the sternal head of the pectoralis major, hypoplasia of the rib cage, and hypoplasia of the upper extremity. In 1841, Sir Alfred Poland described this chest wall anomaly in the Guy's Hospital Gazette while still a medical student based on findings of one cadaver dissection. In his original description, titled "Deficiency of the pectoral muscles," he specifically noted absence of the sternocostal portion of the pectoralis major muscle with an intact clavicular origin, absence of the pectoralis minor, and hypoplastic serratus and external oblique muscles. Poland did not outline the breast hypoplasia or hand deformities in his original description. Poland syndrome can present with ipsilateral involvement of the chest muscles, skin and subcutaneous tissues, bones, and upper extremity. The absence of the sternal head of the pectoralis major muscle is considered the minimal expression of this syndrome. Involvement of adjacent muscles, including the pectoralis minor, serratus, latissimus dorsi, and the external oblique, also has been described. The skin of the area is hypoplastic with a thinned subcutaneous layer and the axillary hair may be absent. The nipple is often smaller and higher in both males and females and the breast is generally hypoplastic in females. Skeletal deformities may involve absence of portions of the ribs or costal cartilages anteriorly. In severe cases, anterior lung herniation may be present. The scapula may be smaller with winging, termed Sprengel deformity. The upper extremity also may be hypoplastic. The upper arm, forearm, and fingers may be shortened, which is termed brachysymphalangism. Simple, complete, or incomplete syndactyly also can be found in patients with Poland syndrome.

Chest radiographic and Computed tomographic findings:

Chest wall abnormalities and determining the presence of the latissimus muscle may require evaluation with CT scan. A chest radiograph may be indicated to determine rib abnormalities or the presence of diaphragmatic hernia. During the examination, note the stage of breast development and status of the latissimus dorsi muscle. The development of the latissimus dorsi muscle can be ascertained with the provocative maneuver of downward strain by the patient with his or her hand on the hip. Furthermore, extraocular muscle motion must be confirmed in excluding an associated Möbius syndrome. Examination of lymph nodes and complete blood cell count should assist with evaluation for associated leukemia and non-Hodgkin lymphoma. Chest wall abnormalities and determining the presence of the latissimus muscle may require evaluation with CT scan.

Other

Breast implants and expanders should not be placed in patients who are too young to maturely accept the responsibility of a foreign body and to have the ability to deal with the potential complications. Patients must be able to tolerate occasional expansion procedures. Integrated port tissue expanders should not be used in young patients because the expander can be deflated in attempting to access the port in a moving target.