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Poland's syndrome? What is it?

Poland's syndrome described in 1841, has an incidence of 1 in 30,000, affects men and women equally and is usually unilateral. It is thought to occur due to disruption of subclavian arterial supply at 6 weeks gestation.

Clinical features, depend on the severity of the expression, however include:

- 1. Absence of pectoralis major and/or latissimus dorsi muscles
- 2. Breast hypoplasia (most common congenital cause of breast hypoplasia)
- 3. Hypoplastic skin, absence of subcutaneous fat and axillary hair
- 4. Pectus excavatum/ carinatum, absence ribs, small scapula, vertebral anomalies
- 5. Congenital hand anomalies i.e.: syndactyly
- 6. Associated with Mobius syndrome and increased incidence of lymphoma

Usually there are no functional concerns with Poland's unless the hand is affected. The main concerns are often breast hypoplasia, absent anterior axillary fold, depression of pectoralis major and rib deformities.

Males usually have chest wall reconstruction during their teenage years to correct clinical deformity. Women often present at the age of puberty and require both chest and breast reconstruction. The type of reconstruction is dependent on the severity.

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Let us answer some questions you may have • What is Poland's Syndrome?

Are Hypertrophic and Keloid scars the same?

Hypertrophic and keloid scars? Are they the same thing and what do we do about them?

Hypertrophic and keloid scars are both due to excessive dermal fibrosis and cutaneous scarring secondary to injury. Men and women are equally affected, more common in African and Asian people, with a peak incidence of between 10 and 30 year olds.

Pathogenesis is unknown however there are a number of theories relating to cytokine dysfunction, tissue tension and wound hypoxia. However that is where the similarity between the two conditions ends.

Hypertrophic scars confine to surgical wound, occur anywhere especially flexor surfaces on limbs, and occur around 4 weeks following injury. These scars are often asymptomatic. They respond well to conservative management, and often mature and resolve over 1 to 2 years. Recurrence is rare.

Keloid scars are more aggressive and invade surrounding tissue beyond the scar. Commonly they occur around sternum, ear, and upper back and deltoid region. They can occur with minor trauma, and usually appear a number of months after the injury. Keloid scars are associated with itch and pain and recurrence is common.

Since they are different conditions the management of them is also different. Hypertrophic scars respond well to massage therapy early. Silicone sheets are also of some benefit. Avoid steroid injections early as they will result contour deformity once the scar matures due to atrophy of tissue below scar. Surgery is rarely required.

Keloid scars require early intervention. Steroid injection and silicone sheets are the main stay of treatment. Steroids are very effective at reducing symptoms of itch and pain. Surgical revision is only offered for those that are resistant to steroid treatment. There is a very high risk of recurrence post-surgery. Radiotherapy is effective but is used as a last resort treatment.

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CHRISTMAS CLOSURE

Please note that WPRS will be closed from Monday 23 rd December 2013 until Sunday 5th January, and will re open on Monday 6th January 2014