



Skin conditions of children and adults with Down syndrome

Part 2 of 2

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There are many features or disorders of the skin observed in individuals with Down syndrome. These range from common conditions such as xerosis (dry skin) and keratosis pilaris to rare disorders such as elastosis perforans serpiginosa. Part 1 of this article was published in *Voice*, December 2010.

5. Skin infections

Skin infections including folliculitis, furuncles, abscesses and secondary impetigo are also common in people with Down syndrome.

Folliculitis is the most common infection, especially in males. Folliculitis is an inflammation or infection of one or more hair follicles (openings in the skin that enclose hair). It can develop on any part of the body, but is most likely to occur on the scalp, face, or parts of the arms, armpits, or legs not usually covered by clothing. Small, yellowish-white blister-like lumps (pustules) surrounded by narrow red rings are usually present with both bacterial folliculitis and fungal folliculitis. Folliculitis develops when bacteria, such as *Staphylococcus*, or a fungus enters the body through a break in the skin near a hair follicle. Scratching the affected area can trap fungus or bacteria under the fingernails and spread the infection to hair follicles on other parts of the body. The bacteria that cause folliculitis are contagious, consequently a person who has folliculitis can infect others who live in the same household. Swabs or skin biopsy may be required to identify the cause of folliculitis. Treatment of recurrent folliculitis is often difficult so antiseptic washes, courses of antibiotics or antifungals or even maintenance antibiotics may be required. People with Down syndrome are also at an increased risk of developing anetoderma, thought secondary to damage to the skin from previous folliculitis. Anetoderma is

a degeneration of the skin in which the skin becomes baggy and wrinkled.

Furuncles (boils) are localised skin infections originating in a gland or hair follicle and characterised by pain, redness, and swelling. Dead tissue deep in the centre of the inflamed area is suddenly extruded, eventually resorbed, or surgically removed. It is important to avoid irritating or squeezing the lesion to prevent spread of the infection. Treatment may include antibiotics, local moist heat, and, when there is definite fluctuation and the hard white core is evident, incision and drainage.

Impetigo is a skin infection that tends primarily to afflict children (school sores). Impetigo is caused by the bacterium *Staphylococcus aureus* (also known as Golden Staph). It tends to develop in areas of the skin that have already been damaged eg through a cut, insect bite, or chickenpox vesicle. The bacteria that cause impetigo are very contagious and can be spread from one part of the body to another by scratching, or contact with a towel, clothing, or stuffed toy. These methods can also pass the infection onto others. Blisters and a golden crust to lesions is characteristic. Antibiotics will be required as well as removal of the affected individual from exposure to others (eg school).

Whilst higher incidences of both scabies and fungal infections have been reported previously in people with Down syndrome, this is thought to be more as

a result of poor hygiene conditions and communal living than any true predisposition. People with Down syndrome do seem predisposed (if they acquire scabies) to a more severe variant known as crusted (Norwegian) scabies. Scabies is an infestation of human skin by an itch mite. The condition is coupled with intense itching, crusting and scaling of the skin, and insect egg burrows that appear as discoloured lines in the affected skin areas. It is highly contagious to anyone in contact with the individual.

6. Vascular skin disorders

The skin has a blood supply (ie delivery of oxygen) in excess of that needed for its own metabolism. Vascular instability is a term used to describe variations in the autonomic nervous system control of this excess blood flow. Vascular instability, in particular acrocyanosis and cutis marmorata, occur with greater frequency in people with Down syndrome than in the general population.

Acrocyanosis occurs because of a decrease in the amount of oxygen delivered to the hands and feet which then turn blue. Decreased blood supply to the affected areas is caused by a constriction of small blood vessels. Acrocyanosis is a painless disorder. The skin still has enough blood supply for its metabolic requirements. No treatment is required.

Cutis marmorata is a pink marble like mottling of the skin caused by exposure to cold temperature. The skin takes on a marbled appearance caused by dilation of small blood vessels. Again the problem is harmless and no treatment is required.

7. Calcinosis cutis

This manifests as small, discrete white lumps under the skin which have a firm to very hard consistency and a chalky appearance. These occur without any symptoms and are most common on the hands and feet. They are caused by the abnormal deposition of calcium salts in the tissues. As many of these lesions eventually heal without scarring, no treatment is necessary.

8. Syringomas of the eyelids

These are not rare in the general population but they appear more common in people with Down syndrome, tending in all populations to be more prevalent amongst females. Syringomas are benign tumours of the sweat glands composed of very small round cysts. Clinically they are smooth skin coloured bumps, approximately the size of a match head and are seen particularly on the lower eyelids. They are usually multiple and symmetrically distributed. They are

asymptomatic, often appear during adolescence, and do not continue to increase in size or become malignant (cancerous). Again, they are essentially a nuisance disorder and may be unnoticeable to the general public unless pointed out. They are fairly simple to remove with gentle ablative techniques (diathermy, cryotherapy or ablative laser) though patient cooperation is essential, otherwise a general anaesthetic would be required. Treatment of any current lesions does not prevent appearance of new ones.

9. Palmoplantar hyperkeratosis

This condition appears to be more common in Down syndrome, and is asymptomatic. The thick skin of the palms and soles is thicker than usual, and often cracks or splits. Moisturiser and topical keratolytics are all that is required.

10. Elastosis perforans serpiginosa

This is a very rare skin disorder occurring also in the general population and with an increased frequency and/or severity in people with Down syndrome (though still very rare). It is a disorder of the connective tissue characterised by ring-shaped groups of asymptomatic rough, scaly, keratotic papules whereby the skin is thickened around a central plug of keratin, overlying an accumulation of elastic tissue. People with Down syndrome appear to be predisposed to developing a more generalised form of the disease. Treatment is difficult and ablative therapies seem most effective (cryotherapy or ablative laser).

References

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