Vitiligo

Vitiligo is an acquired condition where there is patchy loss of melanin from the epidermis, causing areas of pale skin. There may also be loss of melanin in hair follicles. It is usually seen as an autoimmune disease and is associated with other such diseases.

Aetiology

The appearance of vitiligo is due to the loss of functioning melanocytes from the epidermis. The cause remains unclear. Theories about aetiology include:

- **Autoimmune**: destruction of melanocytes by an autoimmune mechanism. T cells have been found to have a significant role.
- **Neurochemical**: destruction of the melanocytes by neurochemical mediators.
- **Autocytotoxic**: destruction of the melanocytes by a metabolic product of melanin.
- **Biochemical**: reactive oxygen species causing melanocyte damage following complex biochemical pathways.
- **Genetic predisposition**.

None of these theories is entirely satisfactory and the truth is likely to be a mixture of all.

Epidemiology

It occurs in about 0.5-2% of the population. It is more obvious - and therefore reported more in dark-skinned races - but no more prevalent. It is also diagnosed more frequently in women, although sex distribution is in fact equal. It affects all age groups but in more than half of cases, onset is before the age of 20.

Risk factors

- Family history of vitiligo.
- Personal or family history of other autoimmune diseases, particularly thyroid disease. Also pernicious anaemia, Addison's disease and diabetes and many other autoimmune conditions.
- History of melanoma and cutaneous T-cell lymphoma.

Possible triggers

These include:

- Emotional stress.
- Childbirth (hormonal changes or stress-related).
- Skin trauma or injury.
- Exposure to certain chemicals (phenolic/catecholic derivatives) although this may be chemical depigmentation rather than vitiligo.
Classification \[^6\]

Vitiligo is classified into the following types.

**Non-segmental vitiligo (NSV)**
- Subtypes are focal, mucosal, acrofacial, generalised, universal.
- White patches are often bilateral and symmetrical.
- Overlaps with other conditions which have known aetiology and are not classified as vitiligo.

**Segmental vitiligo (SV)**
- Subtypes are focal, mucosal, uni-, bi- and pluri-segmental.
- Rapid onset.
- Unilateral.
- Distribution may approximately match a dermatome.
- Involves hair follicle depigmentation.

**Mixed vitiligo (NSV + SV)**
This is rare.

**Unclassified**
This category allows for a period of time of observation, after which a definitive classification can be made.

**Presentation \[^5, 7\]**

The condition can present at any age but most commonly is noticed before the age of 20 years. There may in some cases be itching at onset of new lesions but mostly vitiligo is asymptomatic. However, there may be enormous psychosocial impact, which should not be underestimated \[^4, 8\]. It also carries the risk of associated autoimmune disease.
The patches of vitiligo tend to be clearly circumscribed areas of whiteness. They are especially obvious if the surrounding skin is dark. As they will not tan in sunlight they become more apparent after exposure. Furthermore, because of the loss of melanin they are also more susceptible to sunburn. Lesions are flat and non-scaly. They are often bilateral and symmetrical. At first there are a few clearly defined lesions that may be slightly darker around the perimeter. They increase in size and number, becoming confluent and it may be difficult, if they are extensive, to decide if it is a dark-skinned person with pale patches or a fair-skinned person with pigmented patches. Any part of the body can be affected. Most often it is found on the fingers and wrists, neck, nipples, navel and genitalia, and the skin around the eyes and mouth. It may also be found in body folds such as the groin and axillae, and in sites of skin injury. Where vitiligo occurs in sites of injury or friction, it is known as the Köbner phenomenon. Hair may be white or grey. It is usually in patches on the scalp but can be generalised. Other body hair, including eyebrows and eyelashes, pubic and axillary hair, may also be affected. This is called leukotrichia. The retina may also be affected. Variants include trichrome vitiligo in which there is an area of partial depigmentation as well as the depigmented and normal skin, so that there are three colours. There may be marginal inflammatory vitiligo in which a raised red periphery occurs either at onset or up to a year later. Blue vitiligo may occur with post-inflammatory hyperpigmentation that proceeds to vitiligo.

Investigations

- The diagnosis is generally made clinically.
- Check for evidence of associated disease such as diabetes, pernicious anaemia, thyroid disease and Addison's disease. Consider blood tests for thyroid function and thyroid autoantibodies.
- A Wood's light can be helpful to exclude superficial fungal infections that fluoresce in the ultraviolet light. Not all fungal infections fluoresce and the colour with which they fluoresce also varies. If a Wood's light is shone on areas of depigmentation, the exact margin is more readily seen on fair skin and the lesions appear a bright blue-white.
- Assess the impact on quality of life.
Differential diagnosis[5]

- **Tinea versicolor.** Lesions are dry and slightly scaly when scratched.
- **Piebaldism.** A rare genetic condition. There is often a white forelock, which may be present at birth.
- **Idiopathic guttate hypomelanosis.** Numerous small white macules (measuring 1-5 mm) are distributed symmetrically on the trunk, arms and legs. Lesions have well-defined borders and normal skin markings. It has a different appearance under a Wood's light.
- **Use of potent topical steroids or other types of scarring.**
- **Tuberous sclerosis.**
- **Post-inflammatory hypopigmentation,** following inflammatory skin conditions such as eczema.
- **Pityriasis alba.** This may be a type of eczema or an inflammatory reaction following mild eczema.
- **Leprosy,** especially the tuberculoid variety.
- **Halo naevus.**
- **Lichen sclerosus and atrophicus.** Itchy white patches on the perineum most frequently.
- **Melasma (chloasma).** Hyperpigmentation of the face in women who are pregnant or on the combined oral contraceptive pill.
- **Albinism.** Generalised lack of pigment, including the eyes, present from birth.
- **Morphea.** Localised thickening of the dermis due to excess collagen.

Management[5, 6]

The variety of treatments available suggests that none is totally satisfactory. The response is highly variable between patients. A Cochrane review in 2015 concluded it was not possible to establish which was the best treatment for vitiligo and that further studies are needed[4]. Guidelines were issued by the British Association of Dermatologists in 2008 (currently being updated)[9] and the European Dermatology Forum in 2012[6].

**General measures[7]**

- Protect against sun exposure, as white patches can only burn and cannot tan. Advise about wearing protective clothing, avoiding the sun at peak sun times and use of high-factor sunscreen. High-factor sunscreens can be prescribed on the NHS if endorsed ACBS. On a more cosmetic note, tanning of normal skin makes the patches of vitiligo more apparent.
- Minimise skin injury, as there is an increased likelihood of new white patches in areas of injured skin.
- Monitor for other autoimmune conditions in adults with NSV. Look for symptoms of diabetes, pernicious anaemia, Addison's disease and thyroid disease. Advise people with vitiligo to report symptoms of these conditions. Blood tests for thyroid function and thyroid autoantibodies should be done at diagnosis and then annually thereafter.
- Assess impact on quality of life and presence of psychosocial problems.

Specific treatment is known to be more effective if started early, when the affected area is small, and in childhood[10].

**Camouflage options**

- Cosmetic camouflage creams may be prescribed on the NHS if endorsed ACBS.
- Refer to Changing Faces[11]. This provides education to the patient and advice for the GP about the appropriate cream to prescribe.
- Self-tanning creams.

**Topical corticosteroids**

- These have an anti-inflammatory and immunomodulating effect.
- They can be used in children and adults with limited (less than 10% of the body area) non-segmental vitiligo for a maximum of two months. Topical corticosteroids should not be applied to the face and should not be used in pregnant women.
- Use a potent topical corticosteroid such as mometasone or betamethasone valerate 0.1%, applied once daily. Discontinue after one month if response is good.

**Topical calcineurin inhibitors**

- Tacrolimus and pimecrolimus creams.
- These can be used in adults and children.
They are used for areas of vitiligo on the head and neck. Initial use is for six months but they can be used for longer if effective. They have a better safety profile than topical corticosteroids.

Phototherapies
- **Narrow-band ultraviolet B (NB-UVB) phototherapy:**
  - Effective in widespread NSV.
  - Effective in combination with topical calcineurin inhibitors.
  - Total body treatment is recommended for lesions more than 15-20% of body area.
  - Used in children and adults who have widespread vitiligo, or localised vitiligo which cannot be managed with topical treatments or which is having a significant impact on quality of life.

- **PUVA (psoralen plus UVA radiation):**
  - Oral or topical psoralen followed by exposure to long-wave UV light for a few minutes.
  - It is more effective on the face and trunk.
  - Treatment is twice-weekly for up to two years.

Oral corticosteroids and other immunosuppressants
- Some studies show benefit in fast-spreading vitiligo to arrest progress.
- On the whole, side-effects and risks outweigh benefit and they are not commonly used.

Surgical treatments
- The top layer of vitiligo skin is removed by shaving, dermabrasion or laser and replaced with pigmented skin.
- For areas where there have been no new lesions or spreading of lesions in the previous 12 months, and no Köbner phenomenon (skin lesions on lines of trauma).

Surgical treatment options include:
- Non-cultured melanocyte-keratinocyte cell transplantation.
- Punch grafts.
- Blister grafts.
- Split skin grafts.

Depigmentation therapy
- This is an option for dark-skinned people, who have large areas of skin affected.
- The normal skin is treated to depigment it so that all skin is the same colour.
- A monobenzone ethyl ester cream is applied.
- Treatment takes 1-4 months to work.
- It is usually permanent.

Management in Primary Care
- General measures as above.
- Consider and discuss the option of no treatment where appropriate.
- Offer prescriptions for high-factor sun cream and camouflage creams where appropriate.
- Referral to the Changing Faces camouflage service.[11]
- Give information on the Vitiligo Society for support.[12]
- Consider treatment with a topical corticosteroid if:
  - NSV is localised or limited.
  - The facial area is not involved.
  - The patient is not pregnant.
  - Risks are accepted.
  - No treatment is not an option.
  - The patient is an adult.
Indications for referral

- There is diagnostic doubt.
- For a child affected by vitiligo who requires treatment.
- Segmental vitiligo which requires treatment.
- Treatment is required during pregnancy.
- More than 10% of the body area is affected.
- Significant distress is caused by the condition.
- There is facial involvement and treatment is desired.
- The initial treatment strategy has failed.
- The potential adverse effects of topical corticosteroids are unacceptable.

Where first-line treatment is likely to involve a topical calcineurin inhibitor, referral should be made to a dermatologist.

Consider discussion with a dermatologist by phone about management while waiting for an appointment, particularly if the condition is new or rapidly progressing, or in children.

Further reading & references

- Vitiligo; Primary Care Dermatology Society (PCDS)

5. Vitiligo; NICE CKS, February 2016 (UK access only)
7. Vitiligo; DermNet NZ
10. Guidelines for GPs on the diagnosis and management of vitiligo; The Vitiligo Society 2012
11. Changing Faces
12. Vitiligo Society UK

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