### Comorbidities (action performed for this section: new section)

The similarity between the systemic CI immunophenotype and that of psoriasis has led to the hypothesis of associated comorbidities.[80,159] In HI, arthritis can develop as early as 1 year of age and is not uncommon within the first decade of life.[160–163] A retrospective case–control study of 69 individuals with CI examined various comorbidities, including cardiovascular and rheumatic inflammatory conditions, finding an increased prevalence only for migraine and depression. However, the study's statistical power was limited owing to few events and a relatively young population.[164]

In X-linked ichthyosis (XLI), adults with deletions may be at increased risk for cardiac arrhythmias, particularly atrial fibrillation/flutter, necessitating cardiac function monitoring.[165,166] Both children and adult male patients with XLI-causing deletions may also have an elevated risk of psychiatric and behavioural issues, in particular attention deficit hyperactivity disorder.[167,168]

We therefore recommend screening for comorbidities in all adults with CI, particularly migraine and depression, and for cardiac and psychiatric abnormalities in patients with XLI.

## Specific considerations of congenital ichthyosis

Recommendations (with level of evidence and grade) for the management of CI in the neonatal period and NS are presented in Tables 4–6.

### Specific considerations regarding the management in the neonatal period (action: update)

Clinical presentations at birth requiring special care include collodion baby (CB), neonates with HI, congenital ichthyosiform erythroderma (CIE), NS, EI and ichthyosis prematurity syndrome (IPS). Life-threatening complications may include dehydration from increased transepidermal water loss,[169] infections, electrolyte imbalances, disrupted thermoregulation, metabolic wasting and respiratory distress (particularly in IPS).

#### *Management of collodion baby and harlequin ichthyosis*

Clinical presentations and complications are described in Table S3 (see Supporting Information). Evidence is limited, with mainly review papers, retrospective case series or case reports and only one prospective study.[169–181]

Severity varies widely, from potentially lethal HI to self-healing CB, requiring tailored management.[170]

#### *Management of congenital ichthyosis with other neonatal presentations*

NS and some forms of CIE present at birth with erythroderma and peeling but not in the form of CB.[182–184] Neonates may be admitted to a neonatal intensive care unit for close monitoring, with CB guidelines adjusted based on severity. Early recognition of the underlying cause leads to better treatment and prognosis.[185]

EI presents at birth with blistering and skin loss, often confused with epidermolysis bullosa (EB). Care is similar to EB in neonates,[186–188] focusing on avoiding trauma, heat and adhesives. Key management priorities include wound care to prevent infection, pain relief and addressing dehydration and nutrition.

Babies with IPS are at risk for complications owing to prematurity and life-threatening neonatal asphyxia from aspiration of corneocyte-containing amniotic fluid. Immediate oropharyngeal suction, with or without initial ventilation and intubation, is necessary.[189–191]

### Specific considerations regarding the management of Netherton syndrome (action: update)

Patients with NS experience severe skin inflammation and eczematous lesions requiring specific treatment, along with an increased risk of nonmelanoma skin cancers later in life.

### Skin inflammation and eczema lesions

#### *Topical steroids*

Low-strength topical steroids may be used for a limited period for eczematous lesions, bearing in mind the risk of iatrogenic Cushing syndrome from systemic absorption and severe skin atrophy.[192,193]

#### *Topical calcineurin inhibitors*

Several studies and case reports have shown the efficacy of topical tacrolimus ointment (0.03% or 0.1%) and pimecrolimus cream (1%) (available in some European countries).[194–201]

However, concerns about potential systemic absorption, reported in some cases, suggest that their use should be limited to short-term flare management on small areas. If used longer, serum/plasma drug levels (available for tacrolimus only) should be monitored.[195,198]

#### *Phototherapy*

Short-term efficacy of phototherapy [narrow band ultraviolet (UV)B, psoralen plus UVA or UVA-1] was reported in most case reports.[202–206] However, phototherapy, including UVB, is not recommended for short-term use, and long-term use should especially be avoided owing to the notable risk of skin cancer (see below).

#### *Immunosuppressive drugs and intravenous immunoglobulins*

The use of ciclosporin A was ineffective in two patients and is therefore not recommended.[207]

Systemic immunoglobulins have shown safety and effectiveness in a few patients, but we cannot recommend them owing to limited evidence and lack of long-term data.[208,209]

#### *Biologics*

See new section on biologics in Part one.[5]

#### *Future treatments*

In the future, it is likely that targeted therapy will be available (see the new section on therapeutic approaches in Part one).[5]

Downloaded from https://academic.oup.com/bjd/article/193/1/28/8107307 by guest on 10 October 2025