Table 4 Specific considerations of congenital ichthyosis during the neonatal period: recommendations with level of evidence and grade

Recommendations	Level of evidence	Grade	Type of recommendation	Details of the changes (if applicable)	References
Neonatal period *Management of collodion baby (CB) and harlequin ichthyosis (HI)*					158, 169–181
Admission to neonatal intensive care unit for severe CB	3	D	**MR**	Addition: 'severe' cases of CB	
Involvement of a multidisciplinary team and experienced nursing staff	3	D	UR	NA	
Use of incubator	3	D	**MR/DR**	No systematic use of incubator	
• Decision shared with the neonatal team • Start at 60–80% humidity and decrease every 3–4 days to reach ambient conditions. Be aware that higher humidity may promote the growth of bacteria, such as pseudomonas, or fungal infections, especially candidiasis • Optimal temperature: 32–34 °C. Close monitoring of body temperature • Transfer to an open crib when adequate caloric intake and appropriate weight gain					
Monitoring of nutritional and electrolyte balance • Calculation of intake and output • Caloric supplementation • Oro/nasogastric tube if poor sucking (eclabium) or increased metabolic demands) • Daily checking of body weight	3	D	UR	NA	
Emollients • 3–8 times daily • Sterile occlusive ointments, such as white petrolatum with caution regarding the risk of impaired sweating and infections or water-in-oil emollients • Avoid contamination using latex-free gloves, single-use packets or spoon or tongue depressor to remove emollient	3	D	UR	NA	171, 222
• Avoidance of topical active substances like urea (except on localized area such as palmoplantar areas or constriction bands) (see below), lactic acid, iodine-based antiseptics or silver sulfadiazine. Absolute contraindication for the use of salicylic acid					223–227
Bathing: daily bathing is advisable	4	D	UR	NA	

Adapted from the SIGN50 Guideline Developer's Handbook, NHS Scottish Intercollegiate Guidelines Network, revised edition 2019 (https://www.sign.ac.uk/our-guidelines/sign-50-a-guideline-developers-handbook). DR, deleted recommendations; MR, modified recommendations; NA, not applicable; UR, unchanged recommendations.

should follow country-specific guidelines and the identified bacteria.[119,120] This typically involves intranasal topical antibiotics and bleach baths or soap substitutes containing antiseptic. Serial use of antibiotics is discouraged owing to the risk of bacterial resistance.

Fungal infection should be confirmed by culture and treated topically and/or systemically as appropriate. For all the above situations, it is important to consider the increased risk of systemic absorption of topical therapy, especially in children.

Vaccination (new section)

Although data are limited, there is a risk of severe varicella and herpetic infections in CI, justifying varicella vaccination (mandatory in some countries).[121]

Papillomavirus vaccination is recommended for all patients with NS.

Live and killed vaccines should be administered according to the national vaccination schedule. Patients on immunomodulatory biologics should follow country-specific recommendations for live vaccines.

Growth failure and nutritional deficiency (action: update)

Growth failure is common in children with various forms of CI, especially inflammatory CI, owing to increased metabolic rate, epidermal turnover, chronic inflammation and cutaneous protein loss.[122–125] Children with NS are also at high risk of hypernatraemic dehydration and often have severe failure to thrive with early onset food allergy.[124]

Follow-up in children should include at least annual monitoring of height-for-age and weight-for-age percentiles, along with nutritional assessment, particularly for micronutrients (e.g. vitamin D, selenium, iron and zinc).[126] In severe cases, more frequent monitoring may be necessary, with support from a paediatrician and dietician.[125]

In adults with severe disease, nutritional assessments can be conducted and repeated on a case-by-case basis.

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