

AUTOSOMAL RECESSIVE CONGENITAL ICHTHYOSIS (ARCI)



- Congenital ichthyosis is a genetic disorder of skin cornification that usually presents at birth.
- Characterised by persistently dry, thickened, scaly skin that is associated with itch, skin fragility, redness and sometimes lack of sweating.
- Various different genetic mutations have been found.
- Autosomal recessive congenital ichthyosis (ARCI) is one of the most severe form of congenital ichthyosis.
- Netherton's syndrome combines severe ichthyosis, atopy and hair abnormalities.
- These children are usually born prematurely, may have diarrhoea and failure to thrive.
- They commonly have life-threatening bacterial infections in the first year of life.

- Treatment involves regular topical application of moisturisers to prevent extremely painful fissures, cracks, and infections.
- Lifelong topical or systemic retinoids are usually also needed.

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