

Distribution of Ichthyosis in Children Under 15 Years A Questionnaire Study

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Abstract

Aim - To find out the distribution of Ichthyosis in children under the age of 15

Objectives - It is a questionnaire type study based on causes, signs, symptoms and prevention of Ichthyosis among children.

Background - Ichthyosis is a genetic skin disorder. All types of ichthyosis have dry, thickened, scaly or flaky skin. There are many types of ichthyoses and an exact diagnosis may be difficult. Types of ichthyoses are classified by their appearance and their genetic cause. Treatment of Ichthyosis is by application of cream, emollient oils and retinoids

Reason - To find out about the distribution and create an awareness about ichthyosis.

INTRODUCTION

Ichthyosis is a disorder in which dry or scaly and thickened skin can be seen in the patients. "Ichthy" comes from the Greek word fish. A characteristic feature seen in ichthyosis patients are a scaly appearance in the thickened part of the skin.

Ichthyosis can be of many types like

- **Ichthyosis vulgaris**, where mild scaling and dryness is seen. It is also called X-linked ichthyosis. It is the most common type.
- **Epidemolytic ichthyosis**, where thick and mostly spiny dark scales are seen. They usually follow after trauma
- **Lamellar ichthyosis**, thickened and large plate-like scales are seen.
- **Congenital ichthyosiform erythroderma**, where red skin and fine scales are seen
- **Localized ichthyosis**, where thick or scaly skin is localized to the palms of the hands and soles of the feet.

For a long time, the pathophysiology, mechanisms and underlying genetic defects were unknown, although significant progress has recently been made in the understanding of the molecular basis of human epidermal keratinization processes. Since transglutaminase 1 gene (TGM1) mutations were identified as the cause in lamellar ichthyosis (LI) in 1995 [1,2] Mutations in any of the known causative genes can lead to either NBCIE or LI and candidate genes specific to either NBCIE or LI alone have not yet been identified. [3] The most severe form of this disease spectrum is Harlequin ichthyosis (HI) which is caused by mutations in ABCA12, a putative lipid transport protein of the ATP binding cassette (ABC) family [4,5]. Other features include underdeveloped external ears, nasal hypoplasia, bilateral ectropion with occlusion of the eyes and eclabium. Neonates usually die within the first few days of life from infections and dehydration related complications [6]. The first known report of this is in the diary of Reverend Lover Hart in 1750 [7]. Ichthyosis vulgaris is a common autosomal dominant disorder of

keratinization characterized clinically by scaling, keratosis pilaris, hyperlinearity of the palms, and an association with atopy [8-10]. In some cases, small keratohyaline granules can be resolved by electron microscopy, but these are abnormal in structure and have been described as "crumbly" [11].

MATERIALS AND METHODS

Random families of 58 children under the age of 15 years were chosen in the Chennai population and their parents, mainly mothers were questioned about their child's skin. The symptoms of ichthyosis and other questions about their skin were compiled as a questionnaire and the mothers were asked to fill. Based on the information obtained from them the study was proceeded. The statistics was done and graphs were formed based on the questions answered.

RESULTS

After completion of the filling of the questionnaires, statistics of the answers were done. Most of them did not know about the word 'ichthyosis' but later after they were explained, they were able to correlate.



Fig 1

This graph shows the distribution of ichthyosis in children under 15 years.

There was an overall distribution of 43% .

When they were asked about how long their child has been having the symptoms



Fig:2

Most of the children had the symptoms for only about 1-2 months mostly seasonally. Whereas some had for about 2-4 months and few for more than a year.

When asked whether the lesions were generalized or localized in their child's body, A majority of the answers was that it was localized.

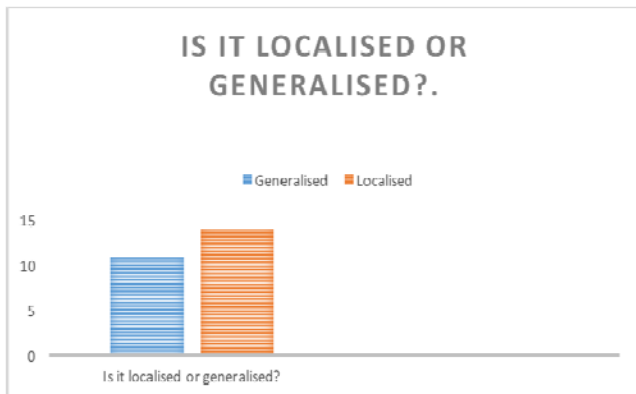


Fig :3

Where was it localized ?? The most common region was the back followed by legs and thighs and lastly in the hands, fingers or in the palm.

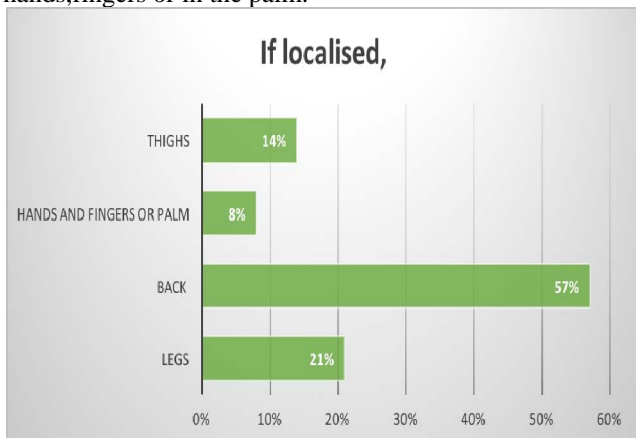


Fig 4

When they were asked whether the symptoms are continuous or do they come and go. Most of them answered that the symptoms come and go



Fig 5

Parents were questioned whether their child has recently come in contact with a person with similar skin and all of them answered that their child has not.



Fig :6

This graph shows if their child has been exposed to something new and only after that the symptoms started.



Fig:7

Parents answered with a 'no' in most cases and in a very few they answered 'yes' and their children were only exposed to new clothing.



Fig :8

Some children had excessive wax buildup in their ears which is a feature of ichthyosis.

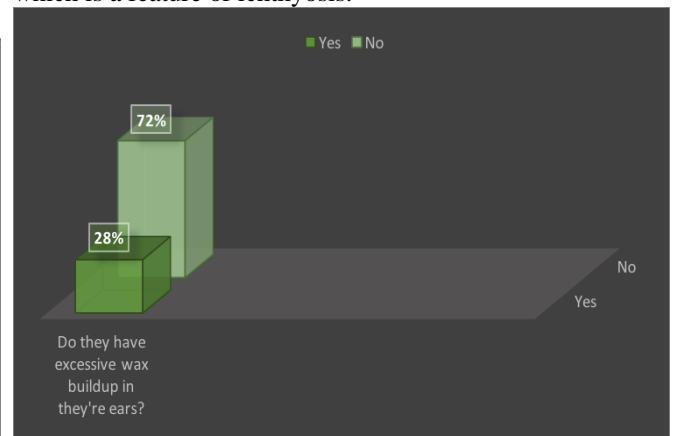


Fig 11

This graph shows if they had any family history with the same symptoms and only one family has previous history of ichthyosis.

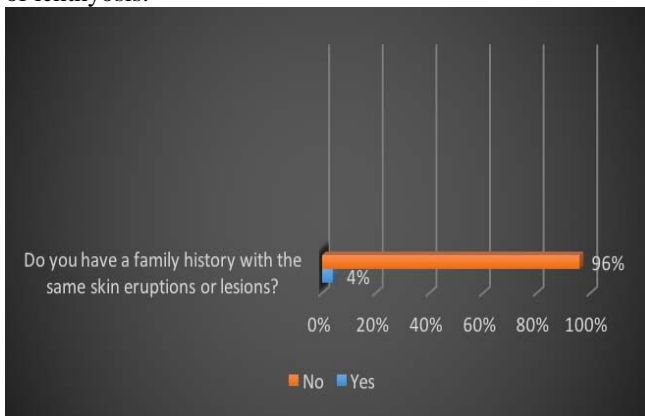


Fig :9

This graph shows whether the child sweats normally, 16% children among the 25 children did not sweat normally.

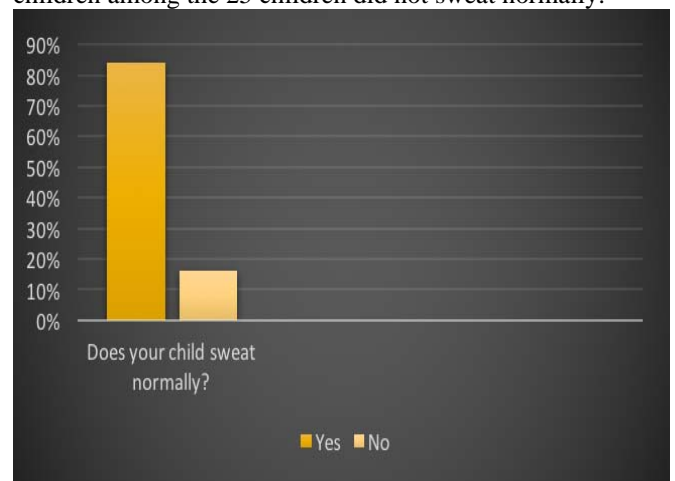


Fig 12

When questioned if their child had a scaly scalp, 4 children did have a scaly scalp which is a characteristic of ichthyosis. When asked if their child had polygonal shaped scales on the skin, 9 children has polygonal shaped scales on their skin.

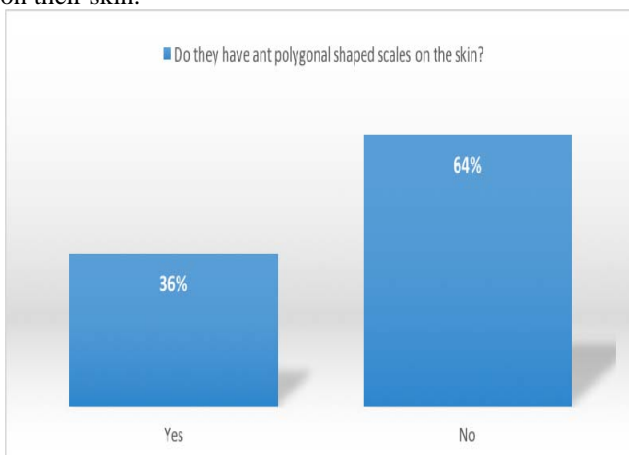


Fig :10

When asked if they consulted any professional about their child's condition, 6 of them consulted a dermatologist only, 4 of them consulted a general physician, 4 of them both a dermatologist and a general physician and 11 of them did not consult a professional at all.

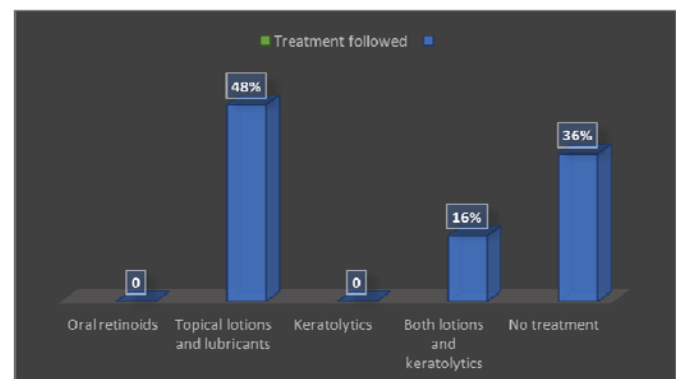


Fig 13

The treatment that was most commonly prescribed was topical lotions and lubricants .

DISCUSSION

Previously, in a study conducted in Spain showed that the calculated distribution of ichthyosis in Romania using the 124 patients for a total number of 16437266 individuals is comparable with the results of 116 individuals in 14408936 French, but with lower estimated distribution [12]. Autosomal dominant ichthyosis vulgaris is characterized by high penetrance and is the most common condition with an estimated distribution of 1:250 to 1:320[13]. It seems to be more frequent in certain areas in India [14]. Its distribution is recorded in literature at 1:6000 [15] in Israel[16] and in Spain 1:4125[17]. Distribution of autosomal recessive lamellar ichthyosis(LI) is estimated at 1:200 000[18].

CONCLUSION

The distribution of ichthyosis in children has increased i.e., 43.1% (25 in 58 children). An awareness about the symptoms, the cause and the treatment should be made among the general public so that even a common man knows what to do when such symptoms arise. Awareness can be made using media especially social media or pamphlets or posters in the city.

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