ECTODERMAL DYSPLASIA ASSOCIATED WITH IMMUNODEFICIENCY

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Background. Mutated genes cause immune system and ectodermal development cause a rare hereditary disorder known as ectodermal dysplasia with immunodeficiency. The intricate relationship between immune system dysfunction and ectodermal abnormalities highlights the difficulty in treating this disorder. **Objective of the study.** To reflect the management methods, clinical presentation, and genetic basis of Ectodermal Dysplasia with Immunodeficiency. **Material and methods.** Literature reviews on ectodermal dysplasia associated with immunodeficiency were utilized, employing inclusion and exclusion criteria, thematic analysis, quality assessment, and ethical considerations across references. Limitations included potential bias and language restrictions. **Results.** The two main genes linked to EDI have been found to be mutated in NEMO and IKBKB. Skin problems, dental anomalies, and scant hair are clinical characteristics. Patients had varying degrees of immunological deficits and were often infected. Immunoglobulin replenishment, skin treatments, and routine dental care are all components of effective management. **Conclusion.** Improving patient outcomes for Ectodermal Dysplasia with Immunodeficiency requires comprehensive care. Genetic testing enables early diagnosis, which enables focused treatment. Multidisciplinary therapy that includes immunological, dental, and dermatological treatment can greatly improve quality of life and lower the consequences. For impacted families, genetic counseling offers invaluable knowledge that helps them comprehend inheritance patterns and make well-informed decisions on family planning. To effectively meet the multifaceted requirements of persons with EDI, therapy and management strategies must be customized and proactive. **Keywords:** ectodermal dysplasia, immunodeficiency, NEMO gene, IKBKB gene, genetic mutations.

CAUSE AND CONSEQUENCES OF PRECOCIOUS PUBERTY

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Background. Precocious puberty (PP) means the appearance of secondary sexual characters before the age of eight years in girls and nine years in boys. There are two types of precocious puberty: central precocious puberty and peripheral precocious puberty. **Objective of the study.** To identify the etiology and consequences of precocious puberty. **Material and methods.** Over 150 publications were viewed and used from databases from online libraries like PubMed, NCBI, USMF library, Webmd and ResearchGate. **Results.** Central precocious puberty is the most common type. It occurs when child's brain releases sex hormones (androgens) too early. Other names for central precocious puberty include gonadotropin-dependent precocious puberty and true precocious puberty. Peripheral precocious puberty occurs because of problems with child's reproductive organs (ovaries or testes) or adrenal glands. Sometimes it results from exposure to hormones in the environment. Linear growth acceleration is one of the important features of early puberty. So, the exact height, weight, growth velocity (cm/year) and BMI should be documented. In females, accurate Tanner staging of the breast should take place, which is particularly challenging in obese or overweight girls to differentiate between adipose tissue and the glandular breast tissue. In males, an orchidometer should be used to determine the testicular volume. **Conclusions.** Precocious puberty is an early onset of puberty and secondary sexual characteristics in children. It is a very difficult diagnosis as the differential ranges from benign variants to serious conditions such as malignancy. This activity reviews the early identification, assessment and management of precocious puberty and highlights the role of healthcare providers in identifying the condition and prompt referral to the pediatric endocrinologist for further management and prevention of complications. **Keywords:** early puberty, sex hormones, adrenal glands.