MCQ's

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Mark the correct answers in the following:

- A- Christ Siemens Touraine syndrome is characterized by -
 - 1- affects males and females
 - 2- affects males only
 - 3- is autosomal dominant
 - 4- is x linked recessive
 - 5- the gene determining the syndrome is mapped to Xq12-q13.1 region of X chromosomal
- B- Cloustonís Syndrome is characterized by -
 - 1- anhydrotic ectodermal dysplasia
 - 2- affects males and females
 - 3- it is autosomal dominant
 - 4- disorganization of hair fibrils
 - 5- biochemical defect in keratin of integumentary system
- C- Clousonís syndrome is characterized by -
 - 1- Partial or total alopecia
 - 2- Coarse fragile hairs
 - 3- Absent or partially absent eye brows
 - 4- Short sparse eye lashes
 - 5- Slow growing thickened nails
- D- Hidrotic or hypohidrotic ectodermal dysplasia is characterized by-
 - 1- Sweat glands may show ductal hyperkeratosis which extends like a pseudo hair
 - 2- Eyes may show strabismus or cataract
 - 3- Buccal leukoplakia in some cases
 - 4- Irregular configuration of hairs
 - 5- Square hair configuration
 - 6- Hair shafts show longitudinal grooving
 - 7- Hair bulbo are dystrophic

- E- Female carriers of x linked recessive ectodermal dysplasia show which of the following markers -
 - 1- Palmar and plantar ridge flattening
 - 2- Low sweat pore count
 - 3- Hypo-dontia of permanent teeth
 - 4- Characteristic dermatoglyphic pattern
 - 5- The skin covering their back may show hypohidrotic area following lines of Blaschko forming iVî shaped area over spine
- F- Cloustonís syndrome may show -
 - 1- reduced dermatoglyphic
 - 2- eccrine poromas
 - 3- palmoplantar hyperkeratosis
 - 4- may get carcinoma of nail bed
 - 5- nails show increased side to side convexity and are prone to infection
- G- Eccrine syringo-fibro-adenoma of Mascaro is -
 - 1- seen in Christ Siemens Touraine syndrome
 - 2- seen in x linked recessive ectodermal dysplasis
 - 3- seen in autosomal dominant hidrotic or hypohidrotic ectodermal dysplasia
 - 4- is a hamartoma or a nevus
 - 5- could represent an induced human papilloma virus epithelial proliferation.
 - 6- Appears as keratotic verrucous or soft nodule with cobble stone arrangement
 - 7- Affects soles and thighs

G-3' 4' 2' 6' 2

E-1' 7' 3' 4' 2

D-1' 7' 3' 4' 2

C-1' 7' 3' 4' 2

B-7' 3' 4' 2