

Polydactyly/Synpolydactyly in Meos and Sunni Muslims of Haryana

Surender Singh¹ and Abhay Singh Yadav²

¹Government P. G. College, Jind, Haryana, India

²Human Genetics Laboratory, Kurukshetra University, Kurukshetra 136 119, Haryana, India

E-mail: surenderkhatkar@gmail.com

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ABSTRACT In the present investigation, nine families belonging to Meos and Sunni Muslims of Haryana were found to be affected with polydactyly and syndactyly. The pedigrees of affected families were analyzed up to a minimum of three generations. An autosomal recessive mode of inheritance of polydactyly/synpolydactyly has been suggested in all these families except one where the possible mode of inheritance seems to be X-linked recessive.

INTRODUCTION

Human limb abnormalities are rare. Polydactyly and synpolydactyly are the congenital abnormalities. Polydactyly is characterized by presence of extra fingers/toes. The commonest type of polydactyly is hexadactyly. Synpolydactyly is characterized by presence of extra digits with webbing. Polydactyly and synpolydactyly are usually inherited as autosomal dominant traits (Bell 1953; Mian 1985; Mathew 1988; Goldstein et al. 1994; Nishikawa et al. 2002). However, in a few cases a recessive mode of inheritance has also been suggested (Gates 1946; Yadav et al. 1991: 92, 94). Preaxial and postaxial polydactyly has been found to be genetically distinct (Temtamy and Mc Kusick 1978) and occasionally coexist in one individual. Postaxial polydactyly has been found to be rare than preaxial polydactyly among Japanese (Satoshi et al. 1991). In addition, the bilateral polydactyly of foot has been found to be the most frequent of foot anomalies (Wantabe et al. 1992). Preaxial polydactyly has been found to be commonest type of hand anomalies (Leung et al. 1982; Oka et al. 1988; Sayeedul et al. 1992). Nishikawa et al. (2002) suggested that bilateral preaxial polydactyly was controlled under autosomal dominant inheritance.

MATERIAL AND METHODS

During the present population genetic survey 200 Meo and 195 Sunni Muslim families were studied. Instances of polydactyly/synpolydactyly were encountered in 9 families belonging to various districts of Haryana. Five families belonging to Meos (Figs. 1-5) and four families to Sunni Muslims (Figs. 6-9) were found affected with polydactyly/synpolydactyly and were analyzed with the help of pedigree charts. A detailed study of each family pedigree was made and possible mode of inheritance of these traits was analyzed. The photographs of the affected individuals were taken for record. The cases of polydactyly and synpolydactyly among the affected families were investigated up to 3rd to 5th generations and were represented in the form of pedigree charts.

RESULTS

Polydactyly and Synpolydactyly among the affected individuals in the pedigrees I to IX have been shown in Tables 1 and 2.

Pedigree I: Individuals III-8 and III-14 were having extra digits in left hand (Fig. 1). The extra digit was preaxial (Table 1). Webbing was present (Table 2). Thus, synpolydactyly was observed. Individual III-8 had a normal son and daughter and two normal sisters. The parents and other relatives of individuals III-8 and III-14 were also normal.

Pedigree II: It revealed a single case of polydactyly. The extra finger was present in both the feet of individual (Fig. 2). The extra digit was

Address for correspondence:

Surender Singh

Assistant Professor,

Government P. G. College,

Jind, Haryana, India

E-mail: surenderkhatkar@gmail.com

Table 1: Types and distribution of extra digits in the polydactyly subjects

| <i>Pedi- gree</i> | <i>Affec- ted indi- vidual</i> | <i>Type-II</i> | | | | | | | | <i>Type-III</i> | | | | | | | |
|-----------------------|--|-----------------|----------|------------------|----------|-----------------|----------|------------------|----------|-----------------|----------|------------------|----------|-----------------|----------|------------------|----------|
| | | <i>Hand</i> | | | | <i>Foot</i> | | | | <i>Hand</i> | | | | <i>Foot</i> | | | |
| | | <i>Preaxial</i> | | <i>Postaxial</i> | | <i>Preaxial</i> | | <i>Postaxial</i> | | <i>Preaxial</i> | | <i>Postaxial</i> | | <i>Preaxial</i> | | <i>Postaxial</i> | |
| | | <i>L</i> | <i>R</i> | <i>L</i> | <i>R</i> | <i>L</i> | <i>R</i> | <i>L</i> | <i>R</i> | <i>L</i> | <i>R</i> | <i>L</i> | <i>R</i> | <i>L</i> | <i>R</i> | <i>L</i> | <i>R</i> |
| I | III-8 | + | - | - | - | - | - | - | - | - | - | - | - | - | - | - | - |
| | III-14 | + | - | - | - | - | - | - | - | - | - | - | - | - | - | - | - |
| II | III-12 | - | - | - | - | - | - | - | - | - | - | - | - | - | - | + | + |
| III | III-3 | - | + | - | - | - | - | - | - | - | - | - | - | - | - | - | - |
| IV | III-6 | + | - | - | - | - | - | - | - | - | - | - | - | - | - | - | - |
| V | II-10 | - | - | - | - | - | - | - | - | - | - | - | - | - | - | + | + |
| VI | III-12 | - | - | - | - | - | - | - | - | - | - | - | - | - | - | - | - |
| | III-3 | - | + | - | - | - | - | - | - | - | - | - | - | - | - | - | - |
| | IV-7 | - | + | - | - | - | - | - | - | - | - | - | - | - | - | - | - |
| VII | IV-9 | - | + | - | - | - | - | - | - | - | - | - | - | - | - | - | - |
| | III-5 | - | - | + | + | - | - | - | - | - | - | - | - | - | - | - | - |
| VIII | II-4 | - | - | - | - | - | - | - | - | - | - | - | - | + | + | - | - |
| IX | III-5 | + | - | - | - | - | - | - | - | - | - | - | - | - | - | - | - |
| | IV-10 | - | + | - | - | - | - | - | - | - | - | - | - | - | - | - | - |

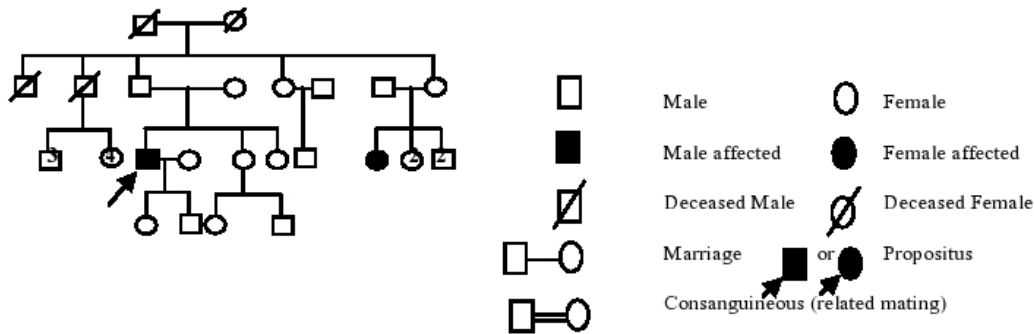


Fig. 1. Pedigree I

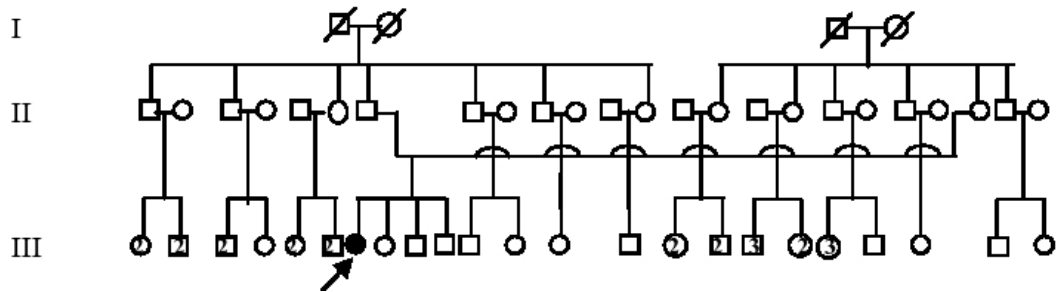


Fig. 2. Pedigree II

postaxial in position (Table 1). Hands of the individual were normal. Webbing was absent (Table 2). Individual III-12 has normal parents and sibs, two brothers and a sister. No other relative of the affected individual was found to be affected with this trait.

Pedigree III: The individual III-3 was having synpolydactyly in the right hand (Fig. 3). The position of extra digit was preaxial (Table 1). Webbing was present (Table 2). The left hand and feet were normal. The propositus has both a daughter and a son. The parents, sister and brother of the affected individual were normal. No other affected individual could be traced either on the maternal or on the paternal side.

Pedigree IV: The individual III-6 had an extra preaxial digit in the left hand (Fig. 4, Table 1). The webbing was absent (Table 2). The right hand and feet of the individual III-6 were nor-

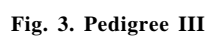
mal. The individuals III-6 had normal parents, sisters and relatives.

Pedigree V: It showed two affected individuals, II-11 and III-12. The propositus individual II-11 had an extra preaxial digit in both the feet while III-12 was having extra preaxial digit in both hands and feet (Table 1). Webbing was also present in feet. Parents and sibs, three brothers and two sisters of II-11 and two brothers and two sisters of III-12, were all normal. Individual II-11 also had two normal daughters and two sons. All relatives of affected individuals were normal.

Pedigree VI: All the affected individuals, III-3, IV-7 and IV-9 had hexadactyly in the right hand only (Figs. 6 and 10). The position of extra digit was preaxial (Table 1). Webbing was absent (Table 2). Left hand and feet of all the affected individuals were normal. III-3 married to a normal individual (III-14) had both the son (IV-

Table 2: Polydactyly and Synpolydactyly among affected subjects in the pedigree under investigation

| Pedigree | Affected individual | Palm | | | | Feet | | | |
|----------|---------------------|-------------------|---|-----------------------------|-----------------------|----------------|---|--------------------------|-----------------------|
| | | Number of fingers | | Fingers involved in webbing | | Number of toes | | Toes involved in webbing | |
| | | L | R | L | R | L | R | L | R |
| I | III-8 | 6 | 5 | | | 5 | 5 | - | - |
| | III-13 | 6 | 5 | | | 5 | - | - | - |
| II | III-12 | 5 | 5 | - | | 6 | 6 | - | - |
| III | III-3 | 5 | 6 | - | thumb and extra digit | 5 | 5 | - | - |
| IV | III-6 | 6 | 5 | - | - | 5 | 5 | - | - |
| V | II-11 | 5 | 6 | - | - | - | - | thumb and extra digit | thumb and extra digit |
| | III-12 | 6 | 6 | - | - | 6 | 6 | thumb and extra digit | thumb and extra digit |
| VI | III-3 | 5 | 6 | - | - | 5 | 5 | - | - |
| | IV-7 | 5 | 6 | - | - | 5 | 5 | - | - |
| | IV-9 | 5 | 6 | - | - | 5 | 5 | - | - |
| VII | III-5 | 6 | 6 | - | - | - | - | - | - |
| VIII | II-4 | 5 | 5 | - | - | 6 | 6 | thumb and extra digit | thumb and extra digit |
| | III-5 | 6 | 6 | - | - | - | - | thumb and extra digit | thumb and extra digit |
| IX | IV-10 | 5 | 6 | - | thumb and extra digit | - | - | - | - |



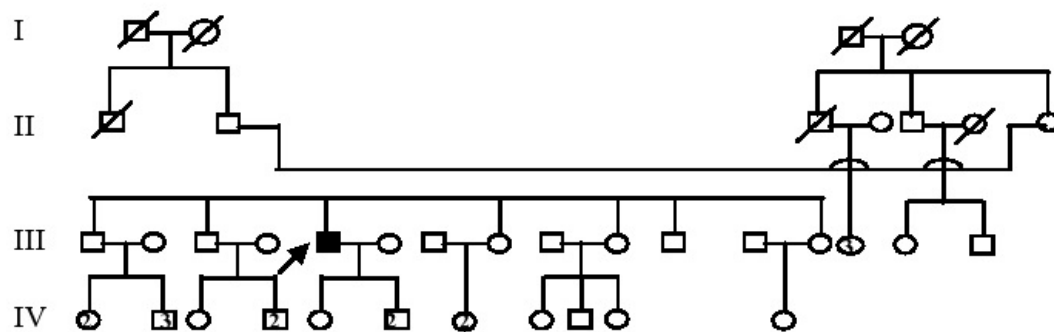


Fig. 7. Pedigree VII

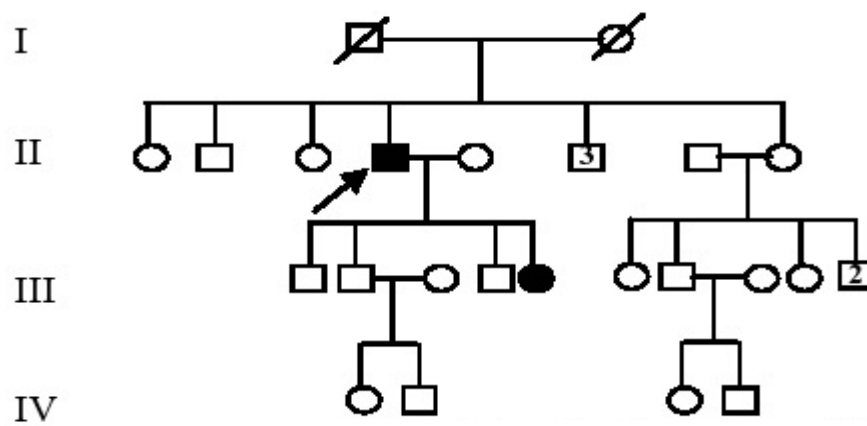


Fig. 8. Pedigree VIII

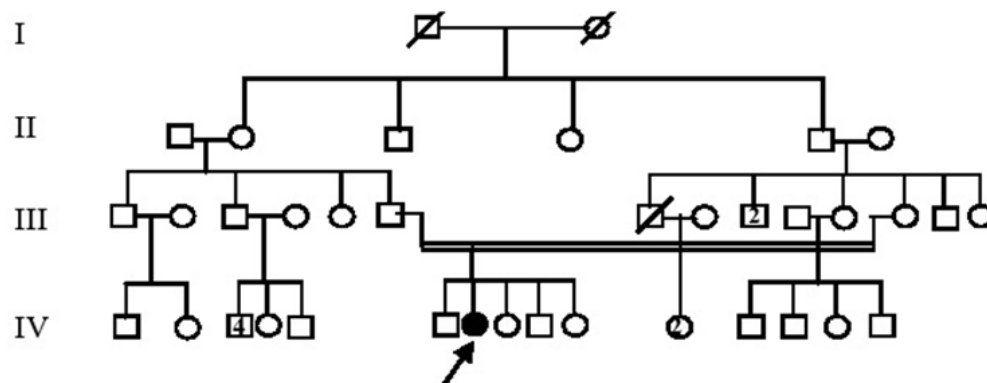


Fig. 9. Pedigree IX



Fig. 10



Fig. 11



Fig. 12



Fig. 13

Fig. 10. Hexadactyly in the right hand of individual III-3 of pedigree VI (Preaxial)
Fig. 11. Hexadactyly in both the hands of individual III-5 of pedigree VII (Postaxial)
Fig. 12. Synpolydactyly in the feet of individual II-4 of pedigree VIII (Preaxial)
Fig. 13. X-Ray photograph of hand confirming Hexadactyly of Gates type II

7) and daughter (IV-9) affected. Individual IV-7 married to a normal individual had two normal daughters and one normal son. No other affected individual was found either on the maternal or the paternal side.

Pedigree VII: Individual III-5 was having postaxial hexadactyly in both the hands (Figs. 7 and 11; Table 1). Webbing of the digits was absent (Table 2). The feet of the individual were normal. The other members of the family were normal.

Pedigree VIII: It revealed two cases of preaxial synpolydactyly in feet. Individual II-4 had an extra finger in right foot while webbing was absent but in left foot extra finger as well as webbing was present (Figs. 8 and 12; Tables 1 and 2). Individual II-4 married to a normal individual and had three normal sons and one affected daughter. Individual III-5 was having an extra digit both in hands and feet which were preaxial in position (Table 1). Webbing was also present between the digits (Table 2).

Pedigree IX: It showed one affected individual IV-10 which depicted preaxial synpolydactyly in the right hand (Figs. 9 and 13; Table 1). Parents and two brothers and two sisters of III-10 were all normal. All relatives of the affected individuals were normal. The webbing was seen between thumb and extra digit (Table 2).

DISCUSSION

During the present investigation, the radiological study of the affected individual was

undertaken, to confirm the different types of polydactyly classified by Gates (1946). Polydactyly in all the affected individuals in different families, except individual III-12 of pedigree II, II-11 of pedigree V and II-4 of pedigree VIII was confined to Gates type II (Figs. 10 and 11). X-ray photograph of the affected individuals confirmed the absence of metacarpal or metatarsal in the extra digit (Fig. 13). However, the affected individual III-12 of pedigree II, II-11 of pedigree V and II-4 of pedigree VIII had one extra digit confirming to Gates type III (Fig. 12).

Further, pedigree analysis revealed that the affected individuals belonging to pedigree II and VII had an extra digit that was postaxial in position. However, the affected individuals belonging to pedigree I, III, IV, V, VI, VIII and IX had an extra digit which was preaxial in position. The frequency of preaxial polydactyly was more than postaxial polydactyly. Since the position of the extra digit among the polydactyl individuals was found to be either preaxial or postaxial, none of the polydactyl individual in all the affected families demonstrated any of the two types of cross polydactyly.

To confirm the possible mode of inheritance for the polydactyly/synpolydactyly, the criteria given by Papp (1990) was followed. The pedigree II, IV and VII had single affected individual with polydactyly. However, pedigree III and IX had single affected individual with synpolydactyly. The appearance of polydactyly/synpolydactyly in these affected families could be sporadic since the trait was confined to single generation only. The possibility of autosomal recessive inheritance cannot be ruled out. In all

these affected families the parents and children of the affected individuals were found to be normal. The autosomal or X-linked dominant mode of inheritance for the above mentioned pedigrees has been ruled out because these do not fit in any of the ten characteristics proposed by Papp (1990). Pedigree I and V involved synpolydactyly. Besides, both the pedigrees had two affected individuals. In both the families, the trait could not express in next generation. The possible reason for this could either be a complete lack of expressivity or incomplete penetrance of defective gene/genes. The trait could also be inherited as autosomal recessive as the parents of the affected cases are asymptomatic and children of the affected parents are themselves normal. The incidence and the severity of the trait are independent of the sex. In pedigree VI the three polydactyly individuals are confined to two different generations. The possible mode of inheritance seems to be X-linked recessive. In pedigree VIII the inheritance appears to be autosomal recessive since there is only one affected child from an affected parent the other parent appeared to be heterozygous.

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