

Letter to the Editor

Effect of Social Factors on the Highest Global Incidence of Congenital Factor XIII Deficiency in Southeast of Iran

Dear Editor,

Factor XIII deficiency (FXIID) is an extremely rare coagulation disorder with an estimated prevalence of 1 in 2 – 3 million in the general population. Although the prevalence of the disease is extremely low, but in an area with high rate of consanguineous marriage such as Sistan and Baluchestan Province is considerable. Sistan and Baluchestan Province has the highest prevalence of the disease worldwide (130 patients per 1 million).¹⁻³

Therefore, this study aimed to assess the role of relatives, ethnicity and residence in this province. Our assessment revealed that consanguinity has a major role in the high prevalence of FXIID in Sistan and Baluchestan Province, southeast of Iran. According to our findings, more than 70 percent of patients had parents with close familial relativity include, uncle girl–cousin, cousin–cousin and cousin–girl of aunt. Only 10 percent of patients' parents had no familial relativity in their parents.

As a result of common cultural features and economic conditions, the high rate of consanguineous marriage in Sistan and Baluchestan Province is similar to other parts of the country. We found that most of the patients (45.9%) were residents of Khash, Zahedan and Saravan cities that have 18.5% and 13.7% of patients respectively. Mirjaveh, Iranshahr, Zabol and Sarbaz cities had 7.8%, 6.8%, 3.9% and 2.4% respectively. These data were obtained from questionnaires. The family tree of patients also showed that the origin of the disease is Khash city and almost all other patients with severe FXIID throughout the province have moved from Khash city to other parts of the province. According to the obtained data more than 90 percent of patients were Baluch and a minority of them was Afghan immigrants (1.5%) and Zaboli (4.5%). This similarity in ethnicity is an auxiliary factor in the rise of consanguinity and therefore the high prevalence of the disease.⁴

It seems that all factors including: ethnicity, residency and consanguinity have an important impact on the highest global prevalence of FXIID in Sistan and Baluchestan Province. This high rate of FXIID, causes high rate of morbidity and mortality in this region. More than 50 patients with FXIID, have experienced CNS bleeding. This life threatening bleeding episode causes high rate of various neurological complications including: behavioral disorders (social dysfunction), developmental disorders, aphasia and hemiplegia. This congenital disorder, also causes a high rate of mortality in the families affected by FXIID. A positive history of death was observed in approximately half of the families with FXIID.⁵⁻⁷

Moreover, lack of a screening test for FXIID in premarital screening program in this region is another contributory factor to the high prevalence of the disorder.

Finally, we concluded that awareness; education, genetic counseling and premarital screening tests are necessary to prevent the spread of such a common occurrence of these bleeding disorders and avoid further spread of the disease in the province.

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