Disease Distribution Manner of people’s body without mutation in DYT1 Gene of Dystonia Disease in Iran

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ABSTRACT

Dystonia is a motional-nervous disorder which is caused by pathologic damages of muscle and led to repetitive and spontaneous movements due to long contraction of muscles which look like ticks and finally directed to paralysis and abnormal gestures of body due to fixed motion which may happen in a muscle or a group of muscles.

INTRODUCTION

This disease is mostly recognized by cramps and torsional motions which involves agonistic and antagonistic muscles. For the first time, this disease was expanded in widespread form of 1 in 9000 Ashkenazi Jews, while in other population it is reported to have a distribution of 1 in 16000 or at least 30%.

Causes:
The environmental reason of the ailment is not acknowledged in some cases. This type is called principal dystonia. In secondary type the environmental cause is identified [32].

Disease Division:

Dystonia is divided into two main categories based on the starting age and distribution in body.

Starting age:
Ailment is divided into two types based on starting age; precocious and serotinous. The precocious type is started at childhood and would undoubtedly have retrieved before the age of twelve. It is usually painful and covers all the body while the serotinous type is accessed at adulthood and mostly covers head and neck.

Body distribution:

Disease is divided into different types based on distribution in body.

Focal: a limited group of muscles are engaged. Its appearance is a type of serotinous. For example, disorders are realized in eyelids so that eyelid opens and closes involuntarily and repeated spasms hinder the eyesight which is called Belfaro spasm.

Regional: a larger group of muscles are engaged. Appearance of the disease is serotinous kind.

Generalized: many parts of the body are involved. It is one of the most painful and debilitating types. Heredity factors are mostly investigated. In addition generalization type is retrieved at the age of five.

Multi focal: in this type of patients will have two symmetrical parts of their body are involved by two arms, engaged.

Hemi-dystonia: disease is distributed totally in one part of the body.

Segmental: two parts of two adjacent parts of body are affianced such as hand and neck [28].
Detection Methods:

Detection methods of this disease are based on blood test, MRI and brain scan or doing genetic test and determining related transfiguration. However, in spite of these methods, there are no certain types of detection method. At present, disease is detected based on symptoms [8].

Treatment:

There is no certain type of treatment for the disease. However, rehabilitation exercises can be caused to reduce muscle disability.

After certain diagnosis of dystonia, first step is using relaxant medicine, in next step of injection; poison is prescribed and caused to be paralyzed for a short time. However, this poison is lost its effect during four to six month and reinjection is needed. During several injections, antibodies are increased onto body and poison is not effective anymore. Injecting Botolinum poison is best type of treatment for focal type of disease. Some patients have been operated under DBS surgical operation. In this operation, an electrode is inserted in cerebral cortex in G lobous Palidous (Palladium) region caused to be invoked Palladium part which certainly many patients will not be responded to this type of surgery [31]. However, this poison is now a primary treatment for focal type. The best method of treatment for this disease is surgery [8].

Primary dystonia:

Some of the patients have mutations in specific genes including DYT1 and DYT6. Its heredity type is Autosomal dominant [28]. Other genes which caused to be generated this disease shows diverse heredity.

Precocious type of this disease is the most painful type caused to cover all muscles of the body and shows mostly generalized type. DYT1 and DYT2 genes are responsible for this ailment [13].

Patients without GAG or DYT1 Mutation

1. The average of beginning age in DYT1 patients

The average age of starting is 13.4 years old which has been doubled in comparison with DYT1⁺. In 37/68% of the patients beginning age is not determined. A disease beginning age is not specified 34/61% of female patients (9 women) and 65/38% of male patients (17 men).

Beginning age before 12 years old:

Among of studied patients, 31/88% of the patients have been appeared the disease before 12 years old that amount of this rate 10/14% (7 people), 13/04% and 7/24% have been appeared before age of 4 and (9 people) before age of 4 to 8 years old (five people) before the age of 12 years old respectively. The proportion of male to female in this range of age has been 0.8%.
1-2 Female patients before the age of 12:
The disease of female patients 38/42 % (10 people) has been displayed before the age of 12. 70 percent of them (7 people), the disease has revealed before the age of 8 and 30% of them (3 people) exposed between the ages of 8 to 12.

3-1 Male patients before the age of 12:
In male patients 30/23 % of them (13 people) has shown the disease before the age of 12 years old. 69/23% of them (9 people) has experienced the disease before the age of 8 and 30/76% (4 people) experience it between the ages of 8 to 12 years old.

4-1 Access over age of 12:
From among the patients 28/98 (20 people) has shown the disease at the age of 12 to 48 which 45% of them are women (9 people) and 55% of them (11 people) are men. It should be added that in women, disease will be more at younger ages and in men, it will be more in older ages.

Distribution parts of body:
Most of the patients who do not have GAG mutation such as DYTI+ show generalized type of disease. Although distribution pattern is not known in 31/88 of patients (22 people), 26/8% of patients (18 people) has shown general type.
general type of distribution:

As was mentioned above 26/8% of the patients (18 people) have shown general distribution, among of them, 61/11% are men (11 people) and 38/88% (7 people) are women. The ratio of male to female is 1 to 0.8. In this type of distribution, different age ranges has been witnessed and the ratio is 24±24. However, beginning age in women has been in childhood and only in one of men it has been at the age of 48. However, distribution in men has been normal. In general type patients, 22/22% (4 people) has the beginning from hand, 22/22% form leg (4 people) and 38/89% from face (7 people). There is no case of disease starting form neck. The patients (4 people) diseases started from right part of body and 5/55% form left part and 16/66% (3 people) from two parts of the body.

Focal Type distribution in patients:

10/14% of the patients showed focal type distribution which from among them 57% are men, 42% (3 people) are women and the ratio of male to female can be reported as 1:0.75. The beginning age in this type of distribution has been 21.5±6.5. As can be seen, it is mostly shown in adulthood and there has not been any precious type. 57% (4 people) has the beginning from hand, 28% form neck (2 people) and 14% from face (1
people). There is no case of disease starting from neck. In 22/22% of the patients (4 people) disease started from right part of body and 5/55% form left part and 16/66% (3 people) from two parts of the body. 42/87% from right part of the body (3 people) and 14/28% (1 person) from left part of the body and 14/28% of them (1 person) has started from two parts of the body and 28/57% (2 people) has started from other patients.

Multi-focal distribution in disease:

After general type, the most distribution has been seen in multi focal form. 14/70% (9 people) have multi focal type which in 66/66% of male (6 people) and 33/33% (3 people) of female shows 1 to 0.5 ratio of male to female. The beginning age of the disease is 14±10. However, in one case disease has been shown earlier and at the age of 4 years old. 44/44% of the patients has shown a start from hand (4 people), 44/44% from leg (4 people) and 11/11% (1 person) from both hand and leg. In addition there has no sign of beginning from face and neck. 33/33% (3 people) has the disease from left and 33/33% from right side and 22/22% from two parts of the body.
Segmental Type:

10/14 % (7 people) has shown segmental type from among which 42/85 % of them are men (3 people) and 57/14% are female (4 people). The ratio of male and female is 1 to 1/35. The only type which has been more in female than male patients occurred in adulthood. Only one patient has segmental type. 42/85 % of the patients (3 people) has started the disease form hand, 28/57% form neck (2 people) and one from leg and one form face.

Hemi-dystonia Distribution:

5/7% of the patients (4 people) has shown hemi-dystonia type of distribution form among which 75% are female (3 people) and 25% are male (one person). So the ratio of male to female is 1 to 3 in this type. One of the patients started the disease form hand, 75% from hand and leg simultaneously. In addition 50% of the patients has the disease from right side (2 people) and 50 % from left (2 people).
Writing Camp Distribution:
1/47% of the patients has a writing camp type of distribution and this kind has been present in only one man at the age of 43 years old (table 4.2.6).
REFERENCES


