Intellectual and Behavior Phenotype of a Sample of Egyptian Patients with Turner Syndrome

Zeinab M. Monir¹, Samira Ismail², Mona O. El Roby², Hassan A. Hussein³, Moshira F. Zaki⁴

Departments of Child Health¹, Clinical Genetics², Cytogenetics³, Biological Anthropology⁴, National Research Centre

ABSTRACT

Turner syndrome (TS) or monosomy X is a genetic disorder in females, approximately half of cases has karyotype 45,X and the other half has mosaicism as isochromosomes of X, ring or partial deletion of X-chromosome. Short stature, infertility with failure of primary and secondary sexual development are the main physical features of Turner syndrome. Impaired psychological adaptation and a characteristic neurocognitive profile has been described in girls with Turner syndrome. The present study included 27 Turner patients, their age range was 6-18 y, with different genetic constitution, we aimed at evaluating the intellectual functions, psychological development and body image among a sample of Egyptian Turner patients, delineating the phenotype / genotype correlation. Patients of the present study were subjected to comprehensive clinical and cytogenetic investigations. Intellectual functions were assessed using Wechsler scales. Psychological evaluation including behavior and self concept was done. According to karyotype, patients of the present study were classified into three groups, classic 45,X (33%), Iso Xq (15%) and mosaic group (42%). Generally, there was downward shift of full intelligence quotient (FIQ), seven cases (26%) of the present study had mental retardation (IQ<70), other ten cases (37%) had educational difficulties (IQ ranged from 70 to 88). Disparity between verbal intelligence quotient and performance intelligence quotient was evident (VIQ>PIQ by 39 digits). Significant increase in violence, withdrawal and psychological troubles was observed. To our knowledge violence has never been reported among Turner patients, it may be a characteristic behavior disorder of Egyptian Turner patients. Poor self esteem and feeling of personal inadequacy was a constant finding among our patients, it was strongly correlated to impaired cognitive functions and behavior disturbance. The present study revealed that isoXq group were the shortest, they had the lowest scores of FIQ, exhibited the highest scores of behavior disturbance. The actual distribution of intelligence quotient among Turner patients need a large scale study lasting for long time through implementation of case registry including all genetics department all over our country to clarify the actual intellectual functions and specify the personality profile of Egyptian Turner patients. Up to date, the cause of impaired neuro-cognitive performance in TS is still uncertain, however the present study raised the possibility of the presence of gene or genes on the short arm of X-chromosome that might influence the cognitive, behavior functions and psychological adjustment. (Egypt J. Neurol. Psychiat. Neurosurg., 2006, 43(1): 23-34)

INTRODUCTION

Turner syndrome (TS) is a genetic disorder in females. It occurs in approximately 1:2000 to 1:5000 live female births. It results from functional absence of the short arm of the second X-chromosome. Approximately half of the cases has karyotype 45,X, and the other half has mosaicism as isochromosome of X, ring or partial deletion of X chromosome¹. TS leads to a variety of physical features, some of which occur in nearly all patients including short stature, primary gonadal failure and infertility with failure of primary and secondary sexual development. Others are less consistent such as webbing of the neck, cubitus valgus, horseshoe kidney and
coarctation of aorta. The loss of the short arm of an X chromosome is associated with short stature. Loss of the long arm, on the other hand, is related to absent ovaries (streak gonads). TS has been estimated to be present in about 1% of all conceptions. One in 15 spontaneous abortions with a chromosomal anomaly has a 45,X karyotype.

It is therefore thought that the 45,X genotype is potentially lethal and that only a minority survive to birth, whether this surviving group have some normal 46,XX cell undetected by routine genetic tests.

Scaffer, first reported a consistent pattern of cognitive strengths and weakness in TS. Temple have emphasized a spatial deficit with verbal skills considered to be normal or strong. Collaer et al., suggested that TS is associated with cognitive alterations.

Many TS females experience impaired psychological adjustment, immaturity and poor self-concept. The present study included 27 TS female patients of different genetic constitution. We aim at evaluating the intellectual functions, behavior adaptation and self concept, accordingly delineating phenotype/genotype correlation

SUBJECTS AND METHODS

The study included 27 TS female patients selected among patients referred to the Human Genetics Clinic, National Research Centre, age range was 6-18 years. They presented because of short stature, delayed puberty and or primary amenorrhea.

All patients were subjected to pedigree construction, full history including age, education, parent's education and occupation, similarly affected sibs or members of family, birth order, and other genetic disorders in the family, with special emphasis on mental and motor developmental history.

Full clinical examination was done for each case. Growth development using anthropometric measurements for weight, height and head circumference was assessed.

Thorough investigations included bone age, Echo cardiography, pelvic sonography and hormonal profile (FSH, LH, Estradiole and progesterone), growth hormone were evaluated after stimulation for some cases.

Cytogenetic analysis was done for every case using G-banding technique and FISH technique. Karyotyping nomenclature was according to the international system for human cytogenetic nomenclatures (ISCN).

Assessment of intellectual functions was done using Wechsler's Scales according to age:

1. Wechsler Preschool and Primary Scale of Intelligence WPPSI, for patients whose age ranges from (4–6 years).
2. Wechsler Intelligence Scale for children WISC, for patients whose age ranges from (6-16 years).
3. Wechsler Adult Intelligence Scale WAIS, for patients whose age is above 16 years.

These scales comprised subtests for verbal and performance intelligence which are the following:

**Verbal subtests**  **Performance subtests**

- Information  **Similarities**
- Comprehension  **Picture completion**
- Digit span  **Object assembly**
- Vocabulary  **Block Design**
- Arithmetic  **Messes**
- Coding

Scoring and scaling of each test was done, verbal intelligence quotient (VIQ), performance intelligence quotient (PIQ), and full intelligence quotient (FIQ) were derived.

**Behavioral Checklist:**

The parental behavioral checklist was translated and developed for Egyptian community by Farag and Ramzy from the behavioral Checklist of the American
Association of Mental Retardation. It includes assessment of violence, withdrawal, hyperactivity and psychological troubles.

* **Self concept (Body Image):** is a self report scale reflecting the attitude of each case towards herself, it was done through questionnaire developed by Shokir, applied individually, it contains 26 items with cut-off 21. Low self esteem was considered at level \( \leq 21 \)\(^{19} \).

**RESULTS**

The study included 27 TS patients, age ranged from 6 to 18 years, mean of age \( 12.8 \pm 3.89 \).

Diagnosis of TS depended on the clinical manifestation of Turner syndrome, patients exhibited short stature, short neck, wide carrying angle, short 4\(^{th}\) metacarpal bone, and primary amenorrhea.

Diagnosis of TS was confirmed by chromosomal analysis using G-banding and FISH techniques, accordingly TS patients were classified into three groups (Table 1 and Figs. 1, 2, 3 & 4).

Anthropometric measurements of studied patients showed that all recruited patients were short (Z score, \(-4.2\) ), patients of IsoXq group were the shortest \((-4.9\) followed by 45,X group \((-4.2\pm1\) and mosaic group \((-4\pm1SD\). ANOVA test revealed no significant statistical difference (Table 2).

The total IQ distribution among the cases illustrated that, mental retardation (IQ<70) represented 26% of the present study (Table 3).

Comparing the cognitive functions (VIQ, PIQ, FIQ) of the cases to those reported of other studies matched with age, we noticed a decrease in values of PIQ and FIQ of our cases, however there was no significant statistical differences (Table 4).

The distribution of cognitive functions (VIQ, PIQ, FIQ) of the studied patients according to genotype groups, showed that the lowest scores of VIQ, PIQ, FIQ were achieved by IsoXq genotype group. However there was no statistically significant difference (Table 5).

Comparing the behavioral disturbances among the patients of the present study to normal controls revealed a significant increase in violence, withdrawal and psychological troubles in TS patients, (\(P<0.05\)). The highest scores of behavioral disturbance were noticed among the IsoXq group (Table 6).

Comparing the means of VIQ, PIQ, FIQ, and behavioral disturbance with body image scores showed that low self esteem was associated with low VIQ, PIQ, FIQ and a significant increase in violence, withdrawal, and psychological troubles (Table 7).

Comparing FIQ in mosaic ring group to classic Turner, we noticed minimal increase of FIQ in mosaic ring group compared to 45,X, however there was no statistical significant difference (Table 8).

**Table 1.** Cytogenetic classification of the studied patients.

<table>
<thead>
<tr>
<th>Genotype</th>
<th>45,X</th>
<th>46,X isoXq</th>
<th>*Mosaic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of cases</td>
<td>9</td>
<td>4</td>
<td>14</td>
</tr>
<tr>
<td>Age range</td>
<td>7-18</td>
<td>10-16</td>
<td>6-18</td>
</tr>
<tr>
<td>Mean of age±SD</td>
<td>13.5±3.8</td>
<td>13.5±2.5</td>
<td>12.2±4.3</td>
</tr>
</tbody>
</table>

* 45X/46,XrX (N=6) 45X/46,XisoXq(N=2)
45X/47,XXX (N =1) 45X/46,XXp- (N =3)
45X/46,XX (N =2)
Fig. (1): Metaphase spread and G-banding Karyotype showing 46,X r(X).
Fig. (2): Metaphase spread and G-banding Karyotype showing 46, X isoXq.
**Fig. (3):** FISH using wcp X showing 46,X r(X).

**Fig. (4):** FISH using wcp X showing 46,X isoXq.
Table 2. Mean Z scores of anthropometric parameters of different genotype groups.

<table>
<thead>
<tr>
<th>Genotype Group</th>
<th>Weight Mean Z score</th>
<th>Height Mean Z score</th>
<th>*H.C Mean Z score</th>
</tr>
</thead>
<tbody>
<tr>
<td>45,x</td>
<td>-2±0.6</td>
<td>-4.2±1.06</td>
<td>-1.8±1.1</td>
</tr>
<tr>
<td>IsoXq</td>
<td>-1.5±0.3</td>
<td>-4.9±1.04</td>
<td>-0.8±0.3</td>
</tr>
<tr>
<td>Mosaic</td>
<td>-2±0.8</td>
<td>-4±1.12</td>
<td>-2±0.8</td>
</tr>
<tr>
<td>Total</td>
<td>-1.9±0.7</td>
<td>-4.2±1.14</td>
<td>-1.8±0.9</td>
</tr>
</tbody>
</table>

*H.C: Head Circumference

Table 3. Total IQ distribution among the studied cases.

<table>
<thead>
<tr>
<th>IQ range</th>
<th>Classification</th>
<th>No of cases</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;110</td>
<td>Above average</td>
<td>4</td>
<td>15</td>
</tr>
<tr>
<td>90-109</td>
<td>Average</td>
<td>6</td>
<td>22</td>
</tr>
<tr>
<td>89-70</td>
<td>Borderline</td>
<td>10</td>
<td>37</td>
</tr>
<tr>
<td>&lt;70</td>
<td>MR*</td>
<td>7</td>
<td>26</td>
</tr>
</tbody>
</table>

MR* = Mental Retardation

Table 4. Comparison between the results of the present study and other studies regarding VIQ, PIQ, and FIQ.

<table>
<thead>
<tr>
<th>No of cases</th>
<th>Mean of age</th>
<th>VIQ Mean</th>
<th>VIQ SD</th>
<th>PIQ Mean</th>
<th>PIQ SD</th>
<th>FIQ Mean</th>
<th>FIQ SD</th>
<th>Difference Mean</th>
<th>Difference SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Money</td>
<td>36</td>
<td>15y</td>
<td>104.6</td>
<td>16.3</td>
<td>87.6</td>
<td>13.3</td>
<td>96.3</td>
<td>14.7</td>
<td></td>
</tr>
<tr>
<td>Garron</td>
<td>30</td>
<td>10y</td>
<td>95.6</td>
<td>17.3</td>
<td>89.1</td>
<td>17.6</td>
<td>91.9</td>
<td>18.3</td>
<td></td>
</tr>
<tr>
<td>Rovet et al.</td>
<td>16</td>
<td>12.2</td>
<td>95.8</td>
<td>19.2</td>
<td>93.6</td>
<td>18.4</td>
<td>94.6</td>
<td>19.1</td>
<td></td>
</tr>
<tr>
<td>Bender et al.</td>
<td>17</td>
<td>13.1</td>
<td>95.4</td>
<td>10.6</td>
<td>91.4</td>
<td>10.4</td>
<td>95.5</td>
<td>18.2</td>
<td></td>
</tr>
<tr>
<td>MacCuly et al.</td>
<td>24</td>
<td>27.1</td>
<td>99.7</td>
<td>11.2</td>
<td>86.9</td>
<td>10.2</td>
<td>93.4</td>
<td>9.3</td>
<td></td>
</tr>
<tr>
<td>Downey et al.</td>
<td>23</td>
<td>12.3</td>
<td>102.7</td>
<td>88.1</td>
<td>95</td>
<td></td>
<td>95</td>
<td>95</td>
<td></td>
</tr>
<tr>
<td>Temple et al.</td>
<td>19</td>
<td>12.8</td>
<td>98.1</td>
<td>28.2</td>
<td>69.7</td>
<td>14.8</td>
<td>84.3</td>
<td>19.3</td>
<td></td>
</tr>
</tbody>
</table>

Table 5. Verbal, Performance & Full IQ distribution according to genotype groups.

<table>
<thead>
<tr>
<th>Group</th>
<th>VIQ Mean</th>
<th>VIQ SD</th>
<th>PIQ Mean</th>
<th>PIQ SD</th>
<th>FIQ Mean</th>
<th>FIQ SD</th>
<th>Difference Mean</th>
<th>Difference SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>45,X</td>
<td>104.3</td>
<td>26.9</td>
<td>72.2</td>
<td>17.7</td>
<td>89.6</td>
<td>18.8</td>
<td>40.6</td>
<td>16.6</td>
</tr>
<tr>
<td>46,XisoXq</td>
<td>79</td>
<td>14</td>
<td>61.2</td>
<td>16.7</td>
<td>68.5</td>
<td>12.3</td>
<td>24.6</td>
<td>19.6</td>
</tr>
<tr>
<td>Mosaic</td>
<td>100.1</td>
<td>30.8</td>
<td>70.7</td>
<td>12.8</td>
<td>85.5</td>
<td>19.9</td>
<td>36.3</td>
<td>28.3</td>
</tr>
<tr>
<td>Total</td>
<td>98.1</td>
<td>28.2</td>
<td>69.7</td>
<td>14.8</td>
<td>84.3</td>
<td>19.3</td>
<td>39</td>
<td>23.6</td>
</tr>
</tbody>
</table>
Table 6. The distribution of behavioral aspects among different groups compared to controls.

<table>
<thead>
<tr>
<th>Genotype</th>
<th>45,X</th>
<th>46,XIsoXq</th>
<th>Mosaic</th>
<th>Total</th>
<th>Control</th>
</tr>
</thead>
<tbody>
<tr>
<td>Behavior</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Violence</td>
<td>3.7±3.5</td>
<td>9.7±4.9</td>
<td>3.2±2.8</td>
<td>4.1±3.8</td>
<td>2.9±2.6*</td>
</tr>
<tr>
<td>Withdrawal</td>
<td>3±4.4</td>
<td>9±3.6</td>
<td>6.6±4.7</td>
<td>5.8±4.8</td>
<td>3.1±2.5*</td>
</tr>
<tr>
<td>Hyperactivity</td>
<td>4.7±3</td>
<td>2.6±3</td>
<td>3.7±4.6</td>
<td>3.8±3.9</td>
<td>3.2±2.6</td>
</tr>
<tr>
<td>Psychol.troub</td>
<td>11.1±4.7</td>
<td>18.6±2.3</td>
<td>10.8±8.5</td>
<td>11.9±7</td>
<td>7.7±4.1*</td>
</tr>
</tbody>
</table>

*P< 0.05

Table 7. Means of VIO, PIQ, FIQ. Violence, Withdrawal, Psychological trouble in cases with body image score.

<table>
<thead>
<tr>
<th>BIS</th>
<th>Violence</th>
<th>Withdrawal</th>
<th>Psych. trouble</th>
<th>VIQ</th>
<th>PIQ</th>
<th>FIQ</th>
</tr>
</thead>
<tbody>
<tr>
<td>LBIS</td>
<td>4.667</td>
<td>7.16</td>
<td>11.33</td>
<td>97.1</td>
<td>71.2</td>
<td>83.9</td>
</tr>
<tr>
<td>NBIS</td>
<td>2.000</td>
<td>-</td>
<td>9.25</td>
<td>116.7</td>
<td>76.7</td>
<td>97.7</td>
</tr>
</tbody>
</table>

BIS = Body Image Score
LBIS = low body image score ≤21
NBIS= normal body image score >21

Table 8. Comparison between 45, X and mosaic ring group according to the values of FIQ.

<table>
<thead>
<tr>
<th>Genotype Group</th>
<th>No. of cases</th>
<th>FIQ</th>
</tr>
</thead>
<tbody>
<tr>
<td>45,X</td>
<td>9</td>
<td>89.6</td>
</tr>
<tr>
<td>45,X/46,XrX</td>
<td>6</td>
<td>91.6</td>
</tr>
</tbody>
</table>

DISCUSSION

Early reports about Turner syndrome (TS) suggested that all affected individuals had general learning disability.  
Bekkar and Van Germound, concluded that TS is linked with a general shift of Full IQ (FIQ) distribution downwards, so that a substantial number of subjects with TS would be functioning intellectually in the normal range. In a large survey by Nilsen and Sillesen, found the prevalence of learning disability in TS to be the same as in the normal population. Ross et al., reported that TS is not typically associated with general mental retardation unlike other common chromosomal aberrations like Down syndrome.

TS patients of our study demonstrated a general shift of FIQ distribution downwards (FIQ = 84.3±19.3). Seven cases out of the 27 cases (26%) suffered mild mental retardation, their FIQ range was 55-67, they need special education as they are out of mainstream schools. Other 10 cases (37%) with FIQ ranged from 70 to 88, they have educational problems and learning difficulties. Temtamy et al., reported 2 cases out of 10 cases, (20%) having mental retardation. Van Dyket et al., found 12 (7%) mentally retarded patients among 174 Turner syndrome patients. It seems to be a great variability in the incidence of mental retardation depending on number of the cases, tools used and culture. The actual distribution of intelligence quotient within Turner syndrome patients is not very well established.
syndrome patients need to be studied on a wider scale, since a limited scale studies actually represent the frequency of IQ scores in its sample not in the Turner population.

Full IQ and Performance IQ among our studied cases were the lowest when compared to those reported by Bender et al.23 and MacCully et al.24. Such observed deficit may reflect the role of poor socioeconomic status of Egyptian TS patients, caused by poor education and occupation of parents with big family size and low income, in addition to the implication of genetic and hormonal factors caused by ovarian failure that begins in utero.

On the other hand, better verbal IQ compared to performance IQ is an associated cognitive alteration in TS patients and is a characteristic neurocognitive profile (98 VS 69.7). It has been consistently documented by other investigators.33-36

However verbal IQ in particular is usually normal, TS patients of the present study have difficulties in verbal subtests, scores of information were low, scores of arithmetic skills and backward digit span were the lowest. Such observation has been previously reported.30,33

Scaffer7 provided the first report of specific cognitive deficit affecting visuo-spatial cognitive skills in individuals of TS patients.

Specific visuo-spatial deficit was believed to account for the disparity between VIQ and PIQ. The wide discrepancy between VIQ and PIQ was observed in the current study VIQ> PIQ by 39 digits. Temple and Carney37, claimed that TS patients have difficulties with certain tests of executive of frontal horn functions. On the other hand, it was thought that altered neural development particularly of parietal or posterior lobes to contribute in cognitive impairment and visuospatial skills.30,38,39

Significant increase of violence, withdrawal and psychological troubles among Turner syndrome patients of the present study was striking finding, withdrawal and psychological troubles have been previously reported among Turner syndrome patients by McCauley et al.40, Kuntsi41 and Collaer7.

On the contrary, violence has never been reported among Turner patients, we think that it may be a characteristic behavioral disorder of Egyptian Turner patients.

On the other hand significant hyperactivity has not been demonstrated by our patients, although it was consistent in previous studies by El-Abd42 and Collaer et al.7.

Behavior changes in TS patients were suggested to result from three main possibilities: early deficiency of ovarian steroids, genetic, or experiential factor9,43. The present study highlights the socioeconomic standard to be the obvious accused factor among the Egyptian Turner patients added to the hormonal and genetic factors.

Feeling of personal inadequacy and inferiority in comparison with other individuals reflected the low self esteem which was a constant finding in the present study, 23 out of 27 (85%) patients exhibited low self concept, it was associated with low Full IQ (FIQ), increased violence, withdrawal and psychological troubles, we agreed with El-Abd et al.42,44, who pointed out that impairment of cognitive tasks may contribute to psychological and emotional developmental problems among TS patients. It was ascertained by decrease of violence, psychological troubles and disappearance of withdrawal among normal self esteem.

However, the current study included 27 TS patients of different genotype groups, the phenotypic, genotypic correlation has been noted among our patients. IsoXq was the striking group whose short stature is manifested by –4.9 SD compared to –4.2 and –4 in 45,X and mosaic groups respectively. This agreed with work done since 1965 by Ferguson-Smith4 and has not been replicated. It showed that loss of short arms of X-chromosome is associated with short stature when compared to other groups.

The lowest scores regarding VIQ, PIQ and FIQ (79, 61.2, 68.5 respectively) have been achieved by TS patients of IsoXq compared to 45,X and mosaic groups (104.3, 72.3, 89.6 and 100.1, 70.7, 85.5) respectively. This is in accordance with William’ study45.
Additionally, our TS patients of IsoXq group demonstrated the highest scores of violence and psychological troubles compared to 45,X and mosaic group.

Our study included 6 mosaic ring group cases, whose FIQ is 2 points more than 45,X. This finding is against Collin et al.\textsuperscript{46} and Kuntsi et al.\textsuperscript{47} they found that, mosaic ring had full IQ lower than 45,X by 11 and 15.5 points respectively. The intellectual function was suggested to be correlated to the size of the active ring and hence was proportionate to the number of genes whose functional disomy affected brain development and functioning.\textsuperscript{47,48} Kamel et al.\textsuperscript{49} studied the intellectual functions of 13 patients of r(X) Turner patients, she concluded that mental status in females with r(X) chromosome is subjected to multiple factors including the presence or absence of (XIST) (X-Inactivation Specific Transcript) on the ring (X) chromosome and the size and the frequency of active r(X) chromosome in addition to genetic and environmental factors.

In the light of specific phenotype-genotype correlation, our study raised to genetic and environmental factors. Implementation of case registration and large scale study including all genetics departments all over our country are highly recommended to delineate the personality profile of Egyptian TS patients; to clarify phenotypic genotypic correlation properly.

The current study emphasized that TS patients have educational problems which need remedial educational programs to cope with their specific learning disability. They are liable to psychological troubles that can be avoided by early intervention, proper management with psychological support.

Up to our knowledge, the cause of impaired neurocognitive performance in TS is still uncertain, studying structural chromosomal abnormality of TS patients may give clue to gene location on X chromosome that influence cognitive and or behavioral development.

\begin{flushright}
\textbf{REFERENCES}
\end{flushright}

46. Collins AL; Cockwell AE; Jacobs PA; Dennis XR(1994): A comparison of the clinical and cytogenetic findings in nine patients with a r(X) cell line and 16( 45, XO) patients. J Med Genet 31: 528-533.

الملخص العربي

القدرات الذهنية والتغيرات السلوكية لعينة من المرضى المصابين بمتلازمة ترنر

إن متلازمة ترنر من الأمراض الوراثية التي تصيب النبات وهي تنشأ نتيجة فقد أحدى كروموسومات X أو نقص X وظيفي للأندرة القصيرة لكروموسم X، وتميز هذه المتلازمة بقصر القامة والعمق بالإضافة إلى عدم التطور الطبيعي لتكوين الندى وملابس البلوغ الأخرى للنبات. إن هذا البحث يشمل 27 مريضة بمتلازمة ترنر ويفيد هذا البحث تقييم القدرات الذهنية والتغيرات السلوكية لبولاء المرضى.

وقد خلص هذا البحث إلى أن التركيب الوراثي بمتلازمة ترنر يشمل عدة أشكال وأن القدرات الذهنية لبولاء المرضى تتميز بالانخفاض معامل النكاء بوجه عام وأن التغيرات السلوكية المصاحبة لهذه العينة من المرضى المصريين تتم بالعفوان وأي سلوك العنف لم يذكر من قبل لمرضى هذه المتلازمة ويدل أن هذا السلوك قد يكون مرتبطة بالمرضى المصريين. وقد لوحظ أن مريضي الوراثي يشعرون بالذبح الواضح لتقديرهم لأنفسهم وهذا التنبؤ مرتبطة بالعفوان معدل النكاء. وبوليسي هذا البحث يعمل تقييم شامل لبولاء المرضى يشمل جميع المراكز البدنية في مصر للوقوف على المعدل الحقيقي لمستوى النكاء ومدى ارتباط سلوك العنف لمرضى هذه المتلازمة من المصريين.