

Validation of the Persian Translation of the TAND Checklist in Children with Tuberous Sclerosis Syndrome

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Abstract

Background: Tuberous Sclerosis Complex is an autosomal dominant heterogeneous disease that mostly affects the skin and nervous system. The goal of this study was to provide a framework for addressing these patients' psychological and behavioral issues, as well as the therapy and management of these aspects.

Methods: As a pilot study for this investigation, 25 patients with Tuberous Sclerosis were assessed. The presence of psychological disorders in these patients was assessed using the TAND (Tuberous Sclerosis Associated Neuropsychiatric Disorders) checklist. The checklist was translated into Persian first, then back into English, and finally verified by the designer of the original checklist. Interviews with the patients' parents took place twice a week with the help of a caregiver, and the checklist was used to analyze the data.

Results: A high correlation was found between the parents' (or caregivers') evaluations of the checklist during the first and second distributions (with a Pearson value of 0.99 and P-value of 0.001). The coefficient of correlation for question eight was -0.76, indicating an inverse correlation. According to the parents' evaluation, the two-stage distribution of this checklist had a strong and respectable reliability with a Cronbach's alpha of 0.79.

Conclusion: Parents and faculty members generally view the reliability and repeatability of all questions in a satisfactory manner as complete and credible from a position of solidarity.

Key Words: Checklist, Neuropsychiatric, Pediatric Neurology, Psychological Disorders, Tuberous Sclerosis.

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1- INTRODUCTION

Tuberous Sclerosis Complex (TSC) is an autosomal dominant multisystem neurocutaneous disorder characterized by cellular hyperplasia and tissue dysplasia, resulting in hamartomatous lesions of the skin and central nervous system (CNS), with involvement of various organs including the kidneys, heart, eyes, and lungs [1]. The prevalence is one in every 6000 live births, mutations in either TSC1 or TSC2 gene that has been identified in 85% of all cases, which act as the main regulator of mTOR (mammalian target of rapamycin), resulting in its increased activity [2]. Angiofibromas, ash-leaf spots, and shagreen patches are a few examples of the cutaneous lesions that are present in 81 to 95 percent of Tuberous Sclerosis patients. Pathological lesions that affect the CNS, such as giant cell astrocytomas and subependymal nodules as well as cortical or subcortical tubers, are frequently accompanied by neurological symptoms like epilepsy and neuropsychiatric disorders [3]. The majority of TSC patients have epilepsy, and at least half also have cognitive impairments and learning disabilities. These conditions are collectively known as TSC Associated Neuropsychiatric Disorders (TAND). One of the main characteristics of TSC is cognitive impairment, which affects 44–65 percent of patients and is frequently seen in children with autism and autistic behaviors like hyperactivity, inattention, and self-injury [4–7]. The prevalence of these behavioral disorders among children with TSC has been estimated to range between 40 and 90 percent in various studies [8]. This study's goal is to review and validate the TAND checklist in the Persian population in light of the high prevalence of these conditions as well as their associated morbidity and mortality. It also aims to develop a workable checklist for use in clinical settings and subsequent

scientific studies of the neuropsychiatric manifestations of TSC.

2- MATERIALS AND METHODS

The TAND (Tuberous Sclerosis Associated Neuropsychiatric Disorders) checklist was used to check for the presence of psychological disorders during the examination of 25 patients with proven Tuberous Sclerosis in the pediatric age range (0-18 years) at Qaem hospital, in Mashhad. In order to use the checklist, it had to first be translated verbatim into Persian and approved by a pediatric neurologist (Appendix 1). After that, it was back translated into English and sent to the checklist's original designer, who confirmed its coordination with the originally designed checklist. The patients' parents or caregivers were ultimately interviewed using a checklist, and again using the same checklist and a different interviewer two weeks later. This checklist was examined and used for the first time in Iran, as a result 25 patients were chosen and a pilot study was carried out (Ethics ID: IR.MUMS.fm.REC.1394.5).

3- RESULTS

This pilot study was conducted on 25 patients with proven Tuberous Sclerosis in Qaem hospital of Mashhad. The age of the children ranged from 1 to 18 years, and in terms of gender, 9 (36%) were girls and 16 (64%) were boys. The transparency and applicability of the checklist was evaluated by 4 pediatric neurologists, wherein 13.64% of the questions were deemed not useful according to one of the professors. No question was indicated as 'not useful' according to more than one professor; so, none of the questions were removed. In total, 96.97% of the questions were deemed necessary according to half or more of the professors. In general, the parents' or caregivers' assessment of the children from the checklist in the first and second execution had a high correlation (with Pearson value of 0.99 and P-value of

0.001). Within the subscales of the checklist, Questions number 1 through 9, with the exception of number 8 had a high correlation coefficient (with all of them being greater than 0.9), and a significant P-value of 0.001 amongst all 8 questions. Thus, among these questions, due to a correlation coefficient of above 0.8 and P-value of less than 0.05, the data was confirmed to have high continuity and similarity among the questions, with a strong positive association. But in the case of Question 8, the correlation coefficient was -0.76 with a P-value of 0.001, which indicates an inverse correlation. In Question 1 related to the children's growth and development (laughing, talking, sitting and standing) and Question 2 which included language, self-care and mobility, the Cronbach's alpha coefficients were 0.77 and 0.79 respectively, which were acceptable values. Question 3 which was related to social behaviors and communicating, and Question 4 related to psychiatric disorders with diagnosis such as autism spectrum disorders (ASD), ADHD, psychotic disorders and schizophrenia, panic and OCD, had Cronbach's alpha coefficients of 0.97 and 0.98, respectively, suggesting strong reliabilities. Question 5 examining IQ and intellectual disability had a Cronbach's

alpha coefficient of 0.25, which is unacceptable in terms of reliability and reproducibility. This can in part be attributed to the absence of a clear relationship between "tendency to have support and evaluation" and the other two subscales; after removing this item, the scales' Cronbach's alpha increased to 0.47. Question 6 which was related to academic skills including reading, writing, spelling and spelling of words and mathematics, and Question 7 which evaluated cognitive skills such as memory, attention, multi-tasking, executive skills, and awareness of time and place, both had Cronbach's alpha values equal to 0.96, suggesting strong reliabilities. Question 8 which evaluated low self-confidence, and high stress level within the family and in the parents had a Cronbach's alpha coefficient of 0.39, regarded as unacceptable. In the evaluation of this question's components, the removal of the "stress in parents" item, improved the alpha value to 0.46 (**Table 1**). The reason for this might be the lack of a clear connection between parental stress and children's communication and challenges. Overall, the parents' evaluation of this checklist suggested a strong and acceptable reliability with a Cronbach's alpha of 0.79.

Table-1: Assessing the correlation and reliability of the TAND checklist in the first and second distributions

Question	First distribution (N = 25)	Second distribution (N = 25)	P-value	cronbach's alpha reliability
Question 1	R = 0.99	R = 0.99	0.001	0.77
Question 2	R = 0.91	R = 0.91	0.001	0.79
Question 3	R = 0.99	R = 0.99	0.001	0.97
Question 4	R = 0.99	R = 0.99	0.001	0.98
Question 5	R = 0.95	R = 0.95	0.001	0.25
Question 6	R = 0.99	R = 0.99	0.001	0.96
Question 7	R = 0.99	R = 0.99	0.001	0.96
Question 8	R = -0.76	R = -0.76	0.001	0.39
TAND	R = 0.99	R = 0.99	0.001	0.79

4- DISCUSSION

TAND was introduced by the neuropsychiatry panel at the 2012 International Consensus Conference on TSC. The main objective of this new term was to define a single structure to describe the multidimensional biopsychosocial manifestations observed in TSC. In order to produce a suitable Persian-translated TAND checklist for use in clinical settings and subsequent scientific investigations, the goal of this study is to review and validate the original TAND checklist. The objective of the current pilot study was to evaluate the TAND checklist's content accuracy as well as its completeness, clarity, usability, and suitability for clinical use. The TAND checklist was improved using the quantitative and qualitative data collected, and the results of the Pearson's correlation test and Cronbach's alpha suggested a high correlation and strong reliability.

The first study conducted to validate the TAND checklist was conducted by Petrus J. de Vries in 2015 [2], and no other study since then has been done in which the families had evaluated this checklist in terms of content and validity. In their study, a total of 20 parents, caretakers, or TSC individuals had participated. All patients enrolled were confirmed as TSC by a definitive criterion, with an average age of 14.25 and were predominantly male. The results of this study showed an alpha value of 0.88 and a P-value of 0.001 in regards to behavioral problems, and an alpha value of 0.75 and P-value of 0.001 in regards to neuropsychological findings. However, the Cronbach's alpha of Question 8 was determined at 0.36, suggesting a weak reliability. In conclusion, this study found that the checklist was a useful tool in determining neuropsychiatric diseases in TSC patients and their subsequent treatment, with the results being similar to the findings of our study.

In this study, the parents' or caregivers' evaluation of the checklist distributed in two stages yielded a high correlation with a Pearson correlation coefficient of 0.99, along with a Cronbach's alpha value of 0.79 suggesting a strong and acceptable reliability. However, in regard to Question 8, similar to the study conducted by Petrus J. de Vries, an alpha value of 0.39 was obtained, which was attributed to the lack of a clear relationship between parents' stress and their child's communication and challenges. In this case, it is possible that the subscales of this question have no relationship with each other. Additionally, other contributing factors such as low self-confidence and problems within family and between parents should also be investigated. In conclusion, and from the perspective of parents and faculty members of solidarity, the reliability and repeatability of all questions are in a satisfactory manner, generally considered to be comprehensive and credible.

The TAND checklist was designed to provide a simple, easy and relatively quick history for clinical teams and families. Originally, this checklist was designed as a communication framework between family specialists in this important but less investigated field. Therefore, it is better to have this checklist translated into other languages so that more patients can be studied and further validation reports based on the translated versions can be obtained. Results of these studies can help improve the quality of the checklist questions and lead to an international consensus on TSC.

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7- AVAILABILITY OF DATA AND MATERIALS

All relevant raw data will be freely available to any researchers wishing to use them for non-commercial purposes, without breaching participant confidentiality.

8- CONFLICT OF INTEREST

None.

9- REFERENCES

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