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Well-being, familial risk, and transfusion interval in thalassemia-affected families: A two-step cluster analysis

Arulmani Thiyagarajan, Bhagvandas M, Kalpana Kosalram, Sudip Bhattacharya[1]

Abstract:

BACKGROUND: Thalassemia is an inherited blood disorder which is one of the common genetic disorders among children that is increasing worldwide.

AIM AND OBJECTIVE: This study aimed to identify clustering patterns among thalassemia-affected families based on their well-being, transfusion interval, and risk factors using two-step cluster analysis (TCA).

METHODS: The study design is a descriptive, analytical cross-sectional study. The study sample consisted of 125 thalassemia children along with either one of the parents who referred to a thalassemia treatment center of a pediatric hospital in Chennai, Tamil Nadu. The parents and children were evaluated using the Ryff Psychological Well-being Scale and Kidscreen Questionnaire respectively. Data were analyzed using SPSS version 16.0 software.

RESULTS: Out of 125 parents, 86 were father (68.8%) and 39 were mother (31.2%). The mean age of parents was 38 years. Similarly, out of 125 thalassemia-affected children, the mean age of children was 13 years. Six clusters were deducted from the TCA. Parents' well-being variable does not have discriminating power to form cluster division. Three cluster formations were meaningful.

CONCLUSION: TCA, in this study, helps in finding the clusters of families with thalassemia-affected children associated with poor well-being and familial risks, which require attention for medical counseling.

Keywords:

Familial risks, thalassemia, transfusion interval, two-step cluster analysis, well-being

Introduction

Thalassemia is a blood inherited disorder caused due to defective hemoglobin synthesis in the red blood cells of the human body. It is one of the common genetic disorders, which affects nearly 200 million people in the world.[1] Thalassemias are inherited conditions carried in the genes and transfer the disorder from parents to children. Carriers of thalassemia show no symptoms, and they might not know that they were carriers.[2] The chance of passing the disease condition to offspring is determined by the carrier status of father and mother. Thalassemia is not a contagious disease.

In many regions of Asia and Africa, consanguineous marriages currently account for approximately 20%–50% of all unions, and preliminary observations indicate that migrants from these areas continue to have marriages with close relatives when resident in North America and Western Europe.[3] Consanguineous

School of Public Health, SRM Institute of Sciences and Technology, Chennai, Tamil Nadu, [1]Department of Community Medicine, Himalayan Institute of Medical Sciences, Dehradun, Uttarakhand, India

Address for correspondence:
Dr. Sudip Bhattacharya, Department of Community Medicine, Himalayan Institute of Medical Sciences, Dehradun, Uttarakhand, India.
E-mail: drsudip81@gmail.com

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marriages are the most important reason for the blood inherited disorders to run among the families.[4]

Worldwide, thalassemia possesses a serious public health problem due to its high prevalence extending from the Mediterranean Basin and parts of Africa.[5] Thalassemia affects approximately 4.4 live births of every 10,000 live births throughout the world.[6] In India, every year, there are 10 thousand children being born with thalassemia which approximately accounts for 10% of the total world incidence of thalassemia-affected children[7] and one in eight of thalassemic carrier lives in India. According to a recent study done in South India, the prevalence of thalassemia ranges between 0.6% and 15%.[8] The nature and the particular treatment of this chronic disease imposes a heavy psychosocial burden on the patients with thalassemia and their families.[9] Serious complications of the disease may lead to poor well-being at any stage of life of the family.

The function of the family mainly depends on the bond between the family members with their emotional health, physical health, and cognitive and social functioning in good terms.[10] Parents having children with chronic disease, have higher responsibilities than their normal counterpart. Experiences and effects of the family members and their reactions toward the children's chronic condition directly affect each other in the family.[11] Its chronic nature of treatment is a permanent reminder to get depressed and makes it impossible to have a normal life.[12]

Health and well-being of his/her parents and their family functioning depend mainly on the health of their children. Therefore, health status between parents and children is strongly dependent. A family is an important source of support for people with chronic disease.[1] Mother of the children with chronic disease has reported higher levels of stress, anxiety, depression, and feelings of isolation than the mother of healthy children.[13] However, chronic disease may strengthen the family unity. Researchers reported that some parents of children with chronic disease felt that the illness functioned as a unifying force and it increased the sensitivity of the family members toward each other.[9] Continuous encouragement from parents, other adult peers and all other important people in their lives, can bring children to the academic task of acquiring intellectual competencies effectively.[14]

To our knowledge, very limited studies are currently available about the clustering pattern among thalassemia-affected families.

The two-step cluster analysis (TCA) procedure is an exploratory tool designed to reveal natural groupings (or clusters) within a dataset that would otherwise not be apparent. The cluster analysis is an explorative analysis that tries to identify structures within the data. Cluster analysis is also called segmentation analysis or taxonomy analysis. More specifically, it tries to identify homogeneous groups of cases, i.e., observations, participants, and respondents. Cluster analysis is used to identify groups of cases if the grouping is not previously known. Because it is explorative, it does not make any assumption which is a specialty of TCA. The different cluster analysis methods that Statistical Package for the Social Sciences Version-16 (SPSS-16) software offers can handle binary, nominal, ordinal, and scale (interval or ratio) data.[15]

Therefore, this study aimed to identify clustering of a comprehensive number of well-being factors in parents of thalassemia-affected children and subsequently to identify groups of thalassemia-affected children with familial risks.

Methods

This cross-sectional study was conducted at the thalassemia treatment center of a pediatric hospital in Chennai, Tamil Nadu, where a separate specialized unit is available to receive these patients and prepared for blood transfusion and administering injections of iron chelation therapy. All pediatric patients diagnosed with thalassemia in this hospital receive medical care. Thalassemia patients between the age groups of 2 and 24 years were included in this study along with either one of their parents.

The sample size was calculated using the formula:

Z^2pq/d^2 [95% confidence interval (CI), prevalence – 4%].[8] Our sample size was 125.

p = prevalence, q = 1-p, d = precision

The Ryff Psychological Well-being Scale was used for measuring the well-being score of the parents with thalassemia patients. Based on the scoring pattern, the categories of well-being have been marked as low, average, and high. Kidscreen Questionnaire was used for assessing the thalassemia children's health status which is validated and pretested tool for assessing the health status of children and young people. Well-being score of 0–7 was considered as low well-being, 8–15 considered as average well-being, and 16–22 considered as high well-being.

Ethical approval from the Institutional Review Board and Research Ethics Committee was obtained. One of the parents of each thalassemia patient signed a consent paper to participate in the study and received an

explanatory form about the study that was attached to the questionnaire. It includes a statement about patients' right to participate or to refuse to participate in the study. Ethical concepts, anonymity, right to withdraw at any time, and respect for opinions and perspectives for children and their parents were taken into consideration in this study. In addition, required permissions were received from relevant authorities in a thalassemia treatment center, Chennai.

Data were collected through a face-to-face interview with each participant and one of his/her parents by the first author. At the start, all questionnaires were prepared, organized, and classified with serial numbers to ensure the availability of the needed information. The patients and their parents were informed about the aim of the study and their participation would be voluntary.

All statistical analyses were performed with Statistical Package for the Social Sciences Version-16 (SPSS-16) software. A TCA was used to identify groups of parents of thalassemia patients with similar well-being attributes, familial risk, and health status of their children. The well-being attributes, marriage type, carrier status, and blood transfusion status of the children were used as input variables in the TCA, together with the sociodemographics such as age, gender, education status, socioeconomic status, and familial risk (carrier status of parents, marriage type, and tribal status)[16,17] The TCA is used here, due to the mixture of categorical and continuous variables. As stated by Norušis, other cluster analysis approaches will not suffice since they rely on either continuous or categorical data (hierarchical clustering) or on a preset number of clusters to be distilled (K-means cluster analysis), whereas the TCA can perform an exploratory cluster analysis using a combination of different types of variables.[18]

Results

One hundred and twenty-five thalassemic children along with either one of their parents were participated in the study. Out of 125, 86 were father (68.8%) and 39 (31.2%) were mother. The mean age of parents was 38 years; out of the 125 parents, 66 were in the age group of 22–37 years, 48 of them were in the age group of 38–53 years, and 11 of them were in the age group of 54–69 years. Out of the 125 children, 81 (64.8%) children were male and 44 (35.2%) were female. The mean age of the children was 13 years. Samples were belonging to the southern part of Tamil Nadu, India.

Due to their disease condition, thalassemia-affected children have to undergo blood transfusion regularly.[19] However, some children might require twice or thrice of blood transfusion for their survival in a month. They have poor health outcome if their well-being score ranges from 0 to 7 which is considered as low well-being.[20]

Cluster analysis was performed with parents' well-being score, marriage type, and carrier status of the parents and transfusion interval of the children. In that, parents' well-being does not play a role [Figure 1] and other variables play a significant role in clustering [Figures 2-4]. Six clusters were deducted from TCA [Table 1].

The "by-variable" importance charts are produced with a separate chart for each cluster. The variables are lined up on the Y-axis, in descending order of importance. The dashed vertical lines mark the critical values for determining the significance of each variable. For a variable to be considered significant, its *t*-statistic must exceed the dashed line in either positive or negative direction. Since the important measures for the variable parents' well-being does not exceed the critical value in the chart, we can conclude that this variable does not contribute to the cluster [Figure 1]. The other variables such as marriage type, carrier status, and transfusion interval are considered significant , as it is *t*-statistic that exceeds the dashed line [Figures 2-4]. The marriage type is playing a significant role in Cluster 1 and Cluster 2. The variable carrier status contributed in all the clusters except the first cluster and the variable transfusion interval contributed to all clusters except the second cluster.

From Table 1, parents' well-being variable does not have discriminating power to form cluster division. Cluster 1 was presented with the nonconsanguineous type of parents' marriage history and had both the parents as a carrier of thalassemia constituting of 13.7% and mother as carrier state ranging 14.8% with average children well-being score.

Cluster 2 was presented with the consanguineous type of parents' marriage and mother as carriers.

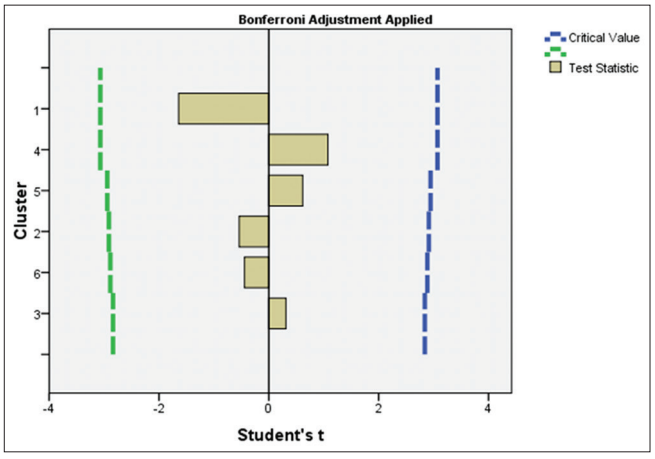

Figure 1: Parents' well-being in cluster importance

Table 1: Cluster of marriage type, carrier state of parents (familial risk), and blood transfusion interval for the children (health outcome)

Components	Cluster1	Cluster2	Cluster3	Cluster4	Cluster5	Cluster6
Cluster distribution (%)	12	17.6	23.2	12	16	19.2
Parents' well-being (%)						
Well-being score (mean)	80.4	82.5	84.7	87.2	85.6	83.1
Children well-being (%)						
Well-being score (mean)	14	15	19	14	20	7
Marriage type (%)						
Consanguineous	0	25.6	24.4	17.4	14	18.6
Nonconsanguineous	38.5	0	20.5	0	20.5	20.5
Carrier status (%)						
Father	0	0	0	0	100	0
Mother	14.8	40.7	0	0	0	44.4
Both	13.7	0	56.9	29.4	0	0
Transfusion interval (%)						
Once	0	0	46.8	0	19.4	33.9
Twice	25.4	37.3	0	23.7	13.6	0
Thrice	0	0	0	25.	0	75

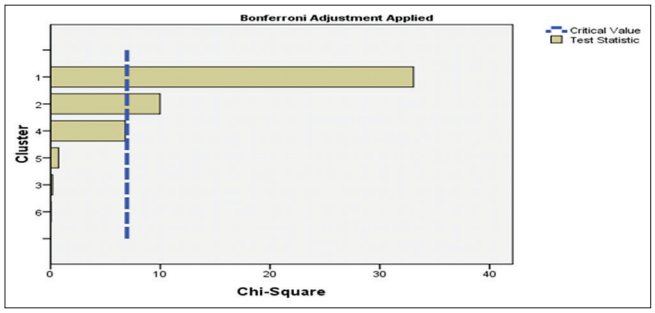

Figure 2: Marriage type in cluster importance

The children had blood transfusion interval of twice per month; children's well-being denotes that they had fair well-being. Cluster 3 was presented with consanguineous marriage and both the parents were being the carrier. Moreover, children had only one blood transfusion in a month and had high well-being score. Cluster 4 was characterized by the parents having consanguineous marriage type as their preference, having both mother and father as the carrier, and their children underwent blood transfusion twice and thrice in a month with average well-being score of their children. Cluster 5 was presented with father alone being the thalassemic carrier and had consanguineous marriage, with children having the blood transfusion of once in a month which showed highest well-being score among all the clusters indicating the healthiest clusters of all. Cluster 6 was characterized with parents having the mixed type of marriage in which 18.6% of parents had consanguineous marriages and 20.5% had the nonconsanguineous marriages. The cluster also exhibited that all the mothers were thalassemic carriers (100%) and their children had low well-being score along with multiple (thrice per month) blood transfusions.

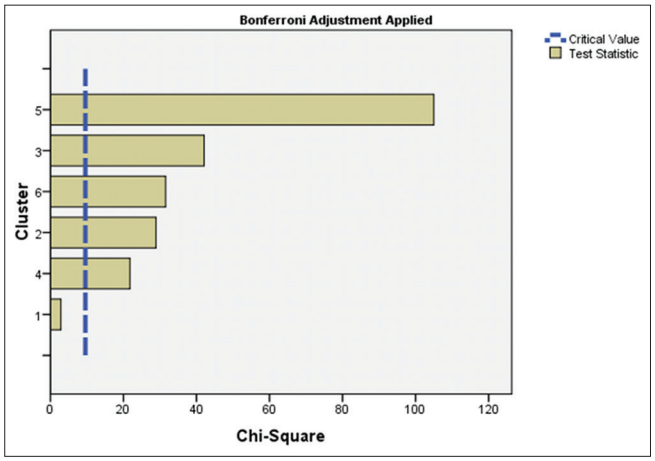

Figure 3: Carrier status of the parents in cluster importance

Discussion

Clustering methods can be applied in various fields which use large datasets, to find the hidden patterns and structure in the dataset with meaningful cluster formation. We used TCA for identifying hidden clusters among thalassemia-affected families which would aim to focus on the risk factors and families that would need medical counseling for sustenance. The familial risk and burden from genetic disorders have always been affecting the child well-being to a greater extent.

TCA reveals six clusters in total. In which, three clusters were meaningful in their structure. Cluster 5 was characterized by father as the thalassemic carrier, history of consanguineous marriage, and children having transfusion interval of only once in a month, which also showed that children's well-being score was high compared to other clusters. This shows that father who has been the main breadwinner for most Indian families

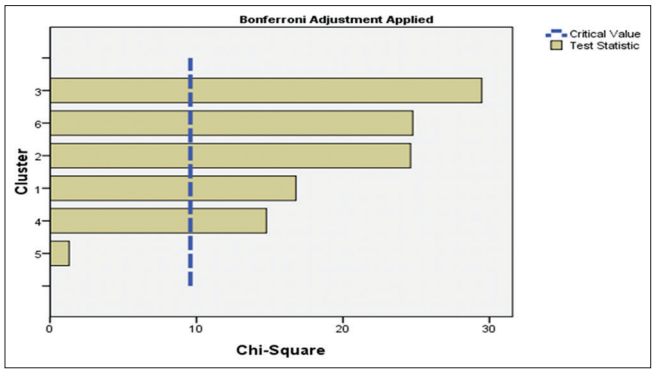

Figure 4: Transfusion interval in cluster importance

scenario[21] cares their children in terms of the following dimensions – caretaker, surety, economic provider, playmate and friend, role model, family head, and also resource support,[22] which make their children to grow in a peaceful life in spite of their disease condition. Cluster 6 has the following characteristics: mother being the thalassemic carrier, history of consanguineous marriage, have their children with blood transfusion interval of thrice in a month predominantly, and also showed low children's well-being score among all the clusters. Female caregivers are much concerned in the future, social support systems, financial difficulty, and the effectiveness of health-care services,[23] which impacted them psychologically and emotionally[24-26] which, in turn, affects their children.[27-30] In Cluster 3, children with both the parents being thalassemic carrier had better health outcome which is evident through children's blood transfusion once in a month with high well-being score. In case of both the parents as the thalassemic carrier, they both give optimal care to the children who improve and regulate the child health better.

From the TCA, we found that parents' well-being does not play a role in cluster distribution compared to other variables. In reality, parents' well-being is very much needed in the children health outcome for effective functioning.[1,31,32]

Unaware of disease status is also huge concern when it comes to familial risk, which put their children 50% of chance to have a disease. Previous studies[33] revealed that there was less knowledge on the detection of carrier status in children and most of the respondents were not heard of the test for detecting thalassemia carrier status. Evidence suggests that it might be a potential factor for children likely to be thalassemic; It is evident from previous studies that the illiteracy and lack of awareness among the parents are also emerged as a contributing factor for this present health condition.[33]

A study of Indian context revealed that by and large, parents have no reservations in sharing information on their affected children with their relatives, and relatives most of them were accepted the risk of being a carrier and even some of them were tested for it, but the communication needs to be improved for all families to accept the risk of having a thalassemia-affected child.[34-37] There is also a need to make the screening more readily available and to motivate high-risk groups through awareness-raising programs.

Thus, for a child to have the better life, both the parents have to get involved in the care of their children. Clustering them in terms of risk groups would help clinicians to identify the high-risk group families. Further studies have to be conducted in different settings and different population groups to validate the results. There might be other risk factors which need to be explored in future studies. The study results should be seen in conjectural way as it is a cross-sectional study.

This study attempts to find the approach for identifying the thalassemia families with high-risk factors, enabling them to have the right counseling and recommended treatment advice for better future sustenance. Different cluster patterns were presented. Parents' well-being has no discriminatory power to divide. Clustering helps physicians to focus on high-risk group of thalassemia-affected families that need attention and medical counseling.

Hence, TCA helps in finding the clusters of thalassemia-affected families associated with poor well-being and familial risks that require immediate attention.

Conclusion

Identifying the cluster among thalassemia-affected families that have poor health outcome and high familial risk would help practitioners to give medical counseling and recommended advice for their normal sustenance.

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Conflicts of interest
There are no conflicts of interest.

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