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Growing up with the families of β-thalassaemia major using an accelerated longitudinal design

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***Background & objectives*: It is difficult for a single investigator to study the psychosocial changes that occur over the life span of an individual affected with a chronic illness like β-thalassaemia major. Therefore, a developmental epidemiological perspective is required to understand the chain of events and problems of psychological nature. We aimed to construct the picture of developmental epidemiology for psychosocial aspects in families of β-thalassaemia major patients attending a tertiary care hospital in north India.**

***Methods*: The accelerated longitudinal design was used. The sample consisted of 100 children with β-thalassaemia and their 150 parents, both groups were subdivided further so that each group represented the continuum of longitudinal course. The sampling was done for a period of 16 months from January 2004 to April 2005.**

***Results*: Overall 54 per cent of children had significant psychopathology. Within the parents groups, 10 per cent had adjustment disorder, 33.3 per cent depressive disorder, and 10 per cent had anxiety disorder and 11 per cent somatoform disorder; 95 per cent of the parents of newly diagnosed children expressed feeling of dazed and shock, fear of death, hopelessness, separation anxiety and problems with their memory and concentration. There was significant difference only in the domain of psychological health in all the three groups of parents with respect to the quality of life. Among children, quality of life improved with their progression of illness. Growing up with β-thalassaemic family was analyzed.**

***Interpretation & conclusions*: The developmental epidemiological perspective was constructed in β -thalassaemic children and their family using an accelerated longitudinal design. Such a design can test the hypothesized aetiological or developmental function of a targeted risk factor within a developmental path and may be used in studying the psychological impact of even other chronic illnesses over the life span of an individual for conceptual and holistic understanding.**

Key words Accelerated longitudinal design - β-thalassaemia - developmental epidemiology - psychosocial aspects - reaction to illness

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In the last decade, several original studies evaluating the psychiatric aspects related to thalassaemia major are reported from India[1-3]. Almost all of these have tried to explore variables like psychiatric co-morbidity, quality of life and coping styles in patients as well as their caregivers by applying simple cross-sectional study design. The results of such studies are often time and situation specific because psychological dysfunction in thalassaemia major assumes the picture of episodic exacerbation as and when the crisis occurs during the course of illness. Since 1960s, with advances in haemato-oncology, median survival in thalassaemia major has increased from 16 to 30 years[4]. Still the stressor of repeated hospitalization with threat of shorter lifespan remains[1]. This stress accrues not only to the patient but impacts on caregivers as well. Inadvertently the sufferings in the family ends only with death of the affected children or the latter just might create another vicious cycle of awaited psychopathology in parents. Single time, simple cross-sectional assessment in such a situation, therefore, might not reflect the comprehensive picture of the sufferings in their family which begin right from the inception of the confirmed diagnosis. Clearly, it is also not feasible for a single investigator to study the longitudinal lines of extensive changes that occur over the life span of an affected individual. In addition, there are always some inherent problems involved in mere cross-sectional and simple longitudinal study design which suggest the desirability of emphasizing the need for more extensive use of a third method. Therefore, a developmental epidemiological perspective is required to understand the chain of events and problems of psychological nature, if age-outcome trajectories over a broad life span need to be assessed.

The concept of developmental epidemiology emerged in the 1980s to bridge the rift between academic (theoretical) and applied (pragmatic) scientific methods so that our concern with the time course of a given disorder, its varying manifestations with development, its precursors and sequelae, and its relation to non-disordered patterns of behaviour becomes clear. Such studies can test the hypothesized aetiological or developmental function of a targeted risk factor within a developmental path, first, by determining if the early antecedent can be changed (the proximal target) and, second, by assessing the impact of changing the proximal antecedent on the distal mental health outcome[5].

In the present article, we aimed to construct a picture of developmental epidemiology in families of β-thalassaemia major. To link together the transition from infancy to childhood and adolescence, we have chosen an accelerated longitudinal design, with cross-sectional single time assessment of child and their respondent parent in families of β-thalassaemia to understand the psychological impact in their families with reference to age-outcome trajectories over a broad age span in short duration. In an accelerated longitudinal design, samples in multiple age groups are included and then the longitudinal data on members of each of them are collected[6].

Material & Methods

Subjects: The universe was the diagnosed and transfusion dependent children with β-thalassaemia major and their parents seeking the treatment from thalassaemia unit in Advance Pediatric Center, Postgraduate Institute of Medical Education & Research (PGIMER), Chandigarh. Three groups of parent (P) and their respective child (C), P1 but no C1, P2 with C2, and P3 with C3 were recruited, each of size 50 separated by 5-7 yr in their life trajectory as if they would represent the real longitudinal continuum. In other words, the entire sample can be hypothetically considered as three family groups spanned by 5 to 7 yr with hypothetical progression in such a way that P3-C3 is an extension of P2-C2 group and later is an extension of P1 but no 'C' group in life trajectory of β-thalassaemia major families. Spanning of 5-7 yr was considered strictly hypothetical to construct developmental epidemiological perspective. Thus total sample consisted of 100 children and 150 parents. However, only single rather than repeated assessment of the entire sample was performed in sequential pattern

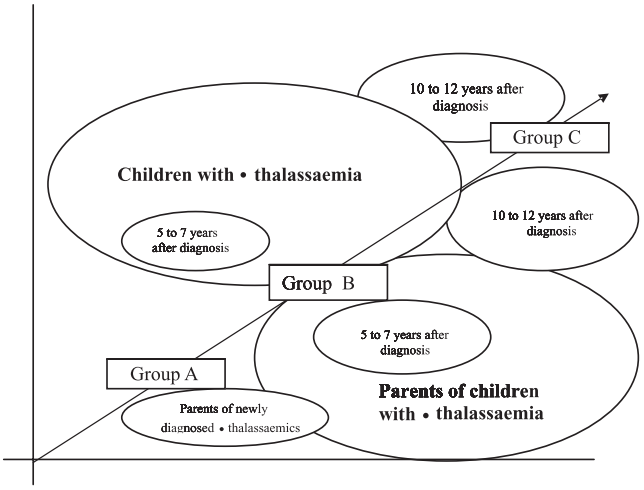

Fig. Explaining the model of methodology used in the study.

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from P1 to P3 and their respective children, adhering to the framework of design. Thus, P1 group of parents were those whose child was immediately confirmed of diagnosis of β-thalassaemia. Their respective children (mean age 16 months) were dropped from analysis because of inability to measure the variables like quality of life and coping in them. P2 and P3 represented the respective parents of C2 and C3 subgroup children (Fig.). The sampling was prospective and only those families were recruited who had accepted our requests and consented for assessments. Overall, 185 families were screened and of them, 35 refused to participate mostly because of time constrains. The rate of acceptability was 81 per cent. Sampling was done for a period of 16 months from January 2004 to April 2005.

Assessments: The procedure and purpose of the study was explained to the participants with the explanation of type of questions they would each be asked. Mothers were preferred as respondents rather than fathers, if both of them were simultaneously present and wanted to participate. Written informed consents from parents were obtained and child's ascent was taken. The administration of respective protocols of assessment for parents and children was completed preferably in one session; each assessment lasting approximately for 2 h. Each patient and the parent were interviewed as per the requirements of the instrument and the instruments were administered in an invariant order. For those parents who were illiterate, investigator read aloud the questions for them. The investigator interviewed the children while parents completed self-report forms. For KIDCOPE[7], children were first prompted to recall a first stressful event by providing an example, and were queried regarding the use of coping strategies in dealing with that situation. For assessing the psychopathology in the parents' group, international classification of diseases (ICD-10)[8] based detailed interviews were taken after psychopathology was screened on comprehensive psychopathology rating scale (CPRS)[9]. At the end of session, the children and their mothers were given a chance to ask questions or raise any concern elicited during the course of the protocol. All the patients continued in the care of their primary treating unit. No change in treatment was done to facilitate intake into the study. Patients diagnosed with psychiatric morbidities were advised to attend psychiatric outpatient services for further management.

Inclusion criteria: *(i)* Either sex (for all groups); *(ii)* Diagnosed cases of β-thalassaemia major (for children group); *(iii)* Respondent parent of the β-thalassaemic children (for parental group); *(iv)* Parents who provided informed consent for the study (for parental group); and *(v)* Children who were aware of their diagnosis and gave assent for evaluation (for children group).

Exclusion criteria: *(i)* Presence of diagnosed chronic physical illness other than β-thalassaemia (for children group); *(ii)* Presence of mental retardation in the child or their respondent parent (for all the groups); *(iii)* Presence of β-thalassaemia in others siblings in family (for all the groups); and *(iv)* History of past psychiatric illness or treatment in child or their respondent parent (for all the groups).

Instruments:

For children

1. Socio-demographic-clinical-profile sheet: This proforma elicits information on age, marital status, occupation, educational status, income, family type, religion, and locality. In addition for children, items for clinical characteristics were added like time since diagnosis, number of transfusion needed every year.

2. Childhood Psychopathology Measuring Scale (CPMS)[10]. This is a comprehensive, reliable and valid instrument adapted and standardized from Child Behavior Checklist (CBCL). It consists of 75 items, which were recorded in question form to make it a semi-structured interview. A cut-off score of 10 of CPMS gave a sensitivity rate of 82 per cent and a specificity of 87 per cent. It is applicable to the children of both the genders in the age range of 4-14 yr.

3. KIDCOPE[7]: It is used to assess children's coping responses to a self-generated stressful situation. The KIDCOPE has two parts, a total of 28 items as separate younger and older version in which the child rates the frequency and efficacy of different coping strategies[7]. The interviewer prompts the child to recall a situation that has bothered him/her in the past month. The one-week correlations ranged from 0.64 to 0.97, and the two weeks scores from 0.68 to 0.90.

4. Malin's Intelligence Scale for Children (MISIC): It is the Hindi version of Wechsler's Adult intelligence Scale-R (WAIS-R)[12]. It has 11 scales, 6 verbal (information, comprehension, arithmetic, similarities, vocabulary and digit span) and 5 performance scales (picture completion, block design, object assembly, codes and mazes).

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5. Quality of Life Scale (PedsQoL)[13]. It is a modular instrument for measuring health related quality of life in children and adolescents from 2-18 yr of age. It is a multidimensional child self-report and parent proxy report scale developed as generic core measures to be integrated with the paediatric quality of life, disease specific module. It consists of physical, emotional, social and scholastic domains with 23 items in total. Both reliability and validity were demonstrated to be equally well.

For parents

1. Socio-demographic sheet
2. Comprehensive Psychopathology Rating Scale (CPRS)[9]. The CPRS covers the full range of psychopathology. This scale is constructed to separate out the reported from observed items, which are 40 and 25 items respectively. Because of the recognized ambiguity of many commonly used psychiatric terms, an explicit description of each item is provided. The scoring system is well accepted and operationally defined. The dimensions used for ranging the scale steps are intensity, frequency and duration of the symptoms. The inter-rater reliability has been found to be satisfactory[9].
3. Ways of coping[14]: It consists of a list of 36 coping strategies used by people to deal with the situations which trouble them. This checklist has high reliability (Cronbach's alpha-0.789). The factor analysis of these strategies grouped them into five coping strategy factors (denial, internalization, externalization, emotional outlet and anger).
4. Wechsler intelligence scale-R: This is a comprehensive test of intelligence applicable to adults from 18 yr onwards that gives separate scores for verbal and performance IQ tests. There are 12 subsets, each of which measures a specific skill or ability. This is most widely used and extensively researched test.
5. Reaction to Illness Questionnaire: It is self-prepared, self-administered instrument in Hindi for assessing immediate reaction of the parents towards the illness of their child. It places emphasise on cognitive, emotional, behavioural social and spiritual domains. The scale has 50 items scored from 0-4 on likert scale. The face and content validity of this instrument was done through 4 different health professionals including two expert psychiatrists, one clinical psychologist and one paediatric haemato-oncology consultant for its semantic potential (unpublished). However, it is yet not properly standardized and any conclusions drawn on its results are tentative and need cautious interpretation.
6. WHO Quality of Life (WHOQOL) – Bref Version[15]: It is a self-administered instrument in Hindi. The WHOQOL scale places emphasis on subjective evaluation of respondent health and living conditions rather than their objective functional status. Four domains of QOL are measured: physical health, psychological health, social relationship and environment. The scale has 26 items, with total score range of 26-130. Its psychometric properties have been found to be comparable to that of the full version WHO QOL - 100. This scale has shown good discriminant validity; content validity, internal consistency and test-retest reliability.

Statistical analysis: All statistical calculations were performed by using the SPSS statistical package (SPSS, Version 10). Descriptive analyses were computed in the terms of means and standard deviations with range for continuous variables and frequency with percentage for ordinal and nominal variables. Non parametric tests were used because the majority of psychometric data were not normally distributed. The results were calculated using Mann-Whitney U test and chi-square test (for categorical variables) and t-test (for continuous variables) to compare two independent groups. Differences with $P<0.05$ were considered significant. Discrete variables were compared between the groups with Fisher's exact test.

Results

No significant intra-class differences were noted in socio-demographic characteristics of parental and their children group (Table I). The mean age of C2 and C3 were 7.5 ± 1.1 and 12.6 ± 1.4 yr respectively; while mean age in the parental groups were 27 ± 7.2, 31 ± 4.4 and 39 ± 5.0 yr respectively. Neurotic traits were seen more in C2 (36%) compared to C3 (30%) group, which might be in conformity to their age; as the children will grow, spontaneous improvement in neurotic traits does occur. Of the 100 children with β-thalassaemia major, 27 were found to have significant psychopathology (CPMS score ≥10). Of these, 34 per cent (17 out of 50) were from C2 and 20 per cent (10 out of 50) were from C3. This might suggest that

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Table I. Socio-demographic characteristics of children of β-thalassaemia major and their parents

Variables	C2 Frequency (%) (n=50)	C3 Frequency (%) (n=50)	P1 Frequency (%) (n=50)	P2 Frequency (%) (n=50)	P3 Frequency (%) (n=50)
Age (yr)					
Mean	7.5	12.6	27.0	31.0	39.0
SD	± 1.1	± 1.4	± 7.2	± 4.4	± 5.0
Sex					
Male	35 (70)	40 (80)	10 (20)	12 (24)	12 (24)
Female	15 (30)	10 (20)	40 (80)	38 (76)	38 (76)
Religion					
Hindu	35 (70)	36 (72)	33 (66)	35 (70)	36 (72)
Non-Hindu	15 (30)	14 (28)	17 (34)	15 (30)	14 (28)
Residence					
Urban	32 (64)	30 (60)	23 (46)	32 (64)	30 (60)
Rural	18 (36)	20 (40)	27 (54)	18 (36)	20 (40)
Family type					
Nuclear	26 (52)	22 (44)	19 (38)	26 (52)	22 (44)
Non-nuclear	24 (48)	28 (56)	31 (62)	24 (48)	28 (56)
Socioeconomic status					
Upper	---NA---	---NA---	21 (42)	14 (28)	11 (22)
Middle			17 (34)	19 (38)	21 (42)
Lower			12 (24)	17 (34)	18 (36)
Neurotic traits					
Present	18 (36)	15 (30)	---NA---	---NA---	---NA---
Absent	35 (70)	32 (64)			
Behavioural problems					
Present	6 (12)	06 (12)	---NA---	---NA---	--NA----
Absent	44 (88)	44 (88)			
Developmental delays					
Present	3 (06)	3 (06)	--NA---	----NA---	---NA---
Absent	47 (94)	47 (94)			
IQ level					
Above 90	48 (96)	49 (98)	---NA----	---NA----	---NA---
Between 7090	2 (4)	1 (2)			
Below 70	0 (00)	0 (00)			
Education					
Literate	---NA-----	---NA----	48 (96)	47 (94)	46 (92)
Illiterate			2 (4)	3 (6)	4 (8)
Occupation					
Employed	----NA----	----NA---	12 (24)	22 (44)	22 (44)
Unemployed			38 (76)	28 (56)	28 (56)

NA, not applicable

the psychopathology develops earlier and severe enough to interfere with growth, development, school performance, or social relationships in pre-adolescent children of β-thalassaemia major. It may be due to the inevitable repeated hospitalizations procedures which often elicit affective interplay in them. It also suggests that as the children would grow into their adolescence, as seen from group A to group B, psychopathology may get remitted to certain extent (14%) may be due to acceptance (of the fact that illness improves through proper care) and adaptation (to necessary procedures as the level of understanding deepens). They (C3) grew up strong. Thus, age may exert pathoplastic effect on psychopathology. Predominantly childhood emotional disorders and oppositional disorder were seen among 17 out of 27, (with CPMS score ≥10). This suggests

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that specific symptoms or distress emerge at particular stages in the developmental process in response to an interaction between the bio-psychosocial processes at work within the individual and the input experienced from the environment. Furthermore, illness-specific factors, such as disfigurement, isolation, and school absenteeism, may disrupt social or identity development.

Of the total 150 parents, 81 were found to have at least one ICD-10 based clinical psychiatric diagnosis (Table II). Within the parent subgroups, 30 per cent (15 of 50) had adjustment disorder exclusively seen in first group (P1) whose child was recently diagnosed, 42 per cent (21 of 50) had depressive disorders in P2 group compared to 18 per cent (9 of 50) in P1 and 38 per cent (19 of 50) in P3. This pattern reflects the likely curvilinear course of depressive psychopathology perhaps cumulating secondary to repetitive burden of hospitalizations and blood transfusions as well as awareness of likelihood of early death approaching to their child. The news that one has a terminal illness takes time to assimilate - to understand all its meanings, grasp all the threats and losses involved. While that process of appraisal is going on, it is difficult to marshal resources and use them effectively. This explains, in part, why adjustment and depressive disorder is more commonly associated with acute than chronic illnesses. The anxiety disorder was seen more in P3 (10%) compared to P1 (2%) and P2 (8%). Somatoform disorder was seen in P2 (8%) and P3 (6%) but was absent in P1. Only one parent had a sleep disorder as a dual diagnosis with depressive disorder. None of the parents had psychotic disorders and all were grounded to reality orientation.

"Reaction to illness" of the parents in three different groups was assessed using self prepared, likert type scale. In P1 group, where the parents were immediately assessed just after disclosure of illness, 94 per cent (47 of 50) had expressed their worries, helplessness, self absorbedness, problems with their memory and concentration; 80 per cent (40 of 50) had expressed denial and feeling of being dazed and shocked; 66 per cent (33 of 50) had attributed the cause of β-thalassaemia major to their bad deeds in the past life compared to 42 per cent (40 of 50) in P2 and 32 per cent (40 of 50) in P3. However, the fear of death was seen comparatively more in P1 (86%) than P2 (70%), in P3, it was again significantly ($P<0.05$) increased to (82%), which can be understood considering course of β-thalassaemia major. In addition, separation anxiety, overprotectiveness, discouraging active coping, spiritual faith and self criticism were found to show attenuation pattern from P1 to P3; while emotional outlet, adaptability with illness and opportunity to serve had shown the incremental pattern of presentation. Irritability, "why me" and feelings of euthanasia remain stable throughout all the groups of parents. Finally, conveying a sense of being valued or respected (44%) was exclusively seen in P3 group. Since the questionnaire is

Table II. Psychiatric morbidity in parents of children with β-thalassaemia major

ICD-10 based psychiatric diagnosis	P1 Frequency (%) N=50	P2 Frequency (%) N=50	P3 Frequency (%) N=50
Present	25 (50)	29 (58)	27 (54)
Depressive disorder	9 (18)	21 (42)	19 (38)
Adjustment disorder	15 (30)	0 (0)	0 (0)
Anxiety disorder	1 (2)	4 (8)	5 (10)
Somatoform disorder	0 (0)	4 (8)	3 (6)
Sleep disorder	0 (0)	0 (0)	1 (2)

Table III. Coping strategies of children with β-thalassaemia major [values are no. (%)]

Coping strategies	Not helped at all		Helped a little		Helped a lot	
	C1	C2	C1	C2	C1	C2
Distraction	3 (6)	3 (6)	47 (94)	47 (94)	00 (0)	0 (0)
Social withdrawal	23 (46)	26 (52)	25 (50)	19 (38)	2 (4)	5 (10)
Cognitive restricting	7 (14)	6 (12)	36 (72)	28 (56)	18 (36)	16 (32)
Self criticism	32 (64)	29 (58)	18 (36)	18 (36)	0 (0)	3 (6)
Blaming others	31 (62)	29 (58)	17 (34)	14 (28)	2 (4)	7 (14)
Problem solving	4 (8)	1 (2)	27 (54)	13 (26)	19 (38)	36 (72)
Emotional regulation	9 (18)	14 (28)	38 (76)	28 (56)	3 (06)	8 (16)
Wishful thinking	14 (28)	4 (8)	25 (50)	31 (62)	11 (22)	15 (30)
Social support	4 (8)	1 (2)	16 (32)	9 (18)	30 (60)	40 (80)
Resignation	33 (66)	33 (66)	16 (32)	13 (26)	1 (2)	4 (8)

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not yet completely standardized, any conclusions drawn on its results are preliminary and tentative.

As far as coping in children was concerned (Table III), emotional regulation was the most frequently (98%) used strategy followed by wishful thinking (86%) and problem solving (80%). Self-criticism was least used (36%) among C2 group. In C3, social support and cognitive restructuring was used by most children (98%) while social withdrawal was used least (36%). In parental groups (Table IV), denial as a coping strategy was used most (78%) while emotional outlet the least (46%). The differences among the three parent groups were significant ($P<0.05$) only in dimensions of internalization, emotional outlet and externalization, whereas they were insignificant in denial and anger categories.

Thus as the children grow, they learn to use more of cognitive restructuring and problem solving. But with age they tend to blame others as well as become more optimistic and their understanding of the illness shapes as family support increases.

In an intra-class comparison of quality of life in children (Table V), both C1 and C2 went through apparently similar phases of physical, emotional, social and school functioning as there was no significant differences observed despite using child and parent report separately. Most of the measures of quality of life in parental group (Table VI) were insignificant; however, psychological health and environment in the family clearly seemed to be disturbed more in P3 compared to P1 and P2 ($P<0.05$). This reflects that as the families of β-thalassaemic children grows from infancy and childhood to adolescence, the parents from P3 group becomes overwhelmingly concerned for the inevitable discomfort that their child would be leaving them soon. This is in contrast to the conventional acceptance of the family system where normally elderly in family dies earlier than the younger members. This psychological blow of impending death clearly causes the deterioration in family environment and psychological health of P3 groups of parents compared to P1 and P2 group wherein the realistic planning for inevitable discomfort been maturely anticipated by facilitating an alliance with treatment team, fearing the worst but yearning for better. In short, P3 parents became more demoralize and hopeless resulting in their poor quality of life.

Like children, adults also vary in their developmental stages and are not homogeneous in

Table IV. Coping strategies of parents of children with β-thalassaemia major

Variables	Group P1 (n=50) Mean ± SD	Group P2 (n=50) Mean ± SD	Group P3 (n=50) Mean ± SD
Denial	3.44 ± 1.2	4.16 ± 2.0	4.76 ± 2.8
Internalization*	4.62 ± 1.5	3.5 ± 2.0	3.28 ± 2.6
Externalization*	1.36 ± 0.8	2.04 ± 1.3	1.72 ± 1.5
Emotional outlet*	0.82 ± 0.5	2.74 ± 0.7	0.98 ± 1.0
Anger	0.36 ± 0.5	0.58 ± 0.5	0.80 ± 0.7
Total	10.80 ± 2.1	11.02 ± 3.9	11.5 ± 6.1

*$P<0.05$ (ANOVA)

Table VI. Intra-class comparison of quality of life in parents

WHO QoL variables	P1 (N=50)	P2 (N=50)	P3 (N=50)
General well being	14.42 ± 1.7	14.54 ± 2.5	14.18 ± 2.3
Physical health	7.02 ± 0.9	7.30 ± 1.2	7.18 ± 1.5
Psychological health*	14.48 ± 1.8	14.36 ± 2.3	13.28 ± 3.3
Social relationship	14.88 ± 2.0	14.76 ± 2.1	14.94 ± 2.2
Environment*	6.98 ± 1.0	6.60 ± 1.3	2.40 ± 1.4
Total	86.57 ± 9.3	86.56 ± 14.1	87.04 ± 12.7

Values are Mean ± SD
*$P<0.05$ (ANOVA)

Table V. Intra-class comparison of quality of life in children

QoL variables	C1			C2	
	Child report (n=50)	Parent report (n=50)	t-value df=49	Child report (n=50)	Parent report (n=50)
Physical functioning	241 ± 75.1	230 ± 83.0	1.091	232 ± 0.91	262 ± 75.1
Emotional functioning	181 ± 47.2	187 ± 48.7	-1.08	187 ± 38.5	179 ± 42.0
Social functioning	169 ± 66.3	177 ± 64.2	-1.38	163 ± 51.3	165 ± 49.4
School functioning	179 ± 49.5	183 ± 51.9	-0..66	167 ± 50.6	172 ± 58.6
Total	763 ± 170.6	778 ± 178.9	-0.72	752 ± 178.4	780 ± 168.5

Values are Mean ± SD

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Table VII. Correlation of childhood psychopathology with age at diagnosis of β-thalassaemic children, their mean IQ, QOL and psychopathology in their parents

Variables	CPMS total score	Age at diagnosis	Mean IQ	CPRS total score	WHO-QOL total score
CPMS total score	1.00	-0.15	-0.05	-0.01	-0.25*
Age at diagnosis	-0.15	1.00	-0.10	-0.05	0.23*
Mean IQ	-0.05	-0.10	1.00	0.01	0.09
CPRS total score	-0.01	-0.05	0.01	1.00	-0.36*
WHO QOL (parents) Total score	-0.25*	0.23*	0.09	-0.36	1.00

Pearson's correlation *$P<0.05$

their ability to cope. Many adults have limited abilities to abstract and to understand illness as seen from developmental epidemiological framework of our study. Correlation of childhood psychopathology with age at diagnosis of β-thalassaemic children, their mean IQ, QOL and psychopathology in their parents is shown in Table VII. Clearly, childhood psychopathology was negatively but significantly ($P<0.05$) correlated with their quality of life. Age at diagnosis and IQ levels were not significant. Thus more the psychopathology poorer is the quality of life in parents and understandably served as predictors for one another.

Discussion

Most studies about psychosocial aspects of β-thalassaemia involved the subjects of mixed duration at study entry[1,2.16-28] and thus are not directly comparable to our study. We considered the accelerated longitudinal design to understand how the families of β-thalassaemia major move on with the time span of about 10 to 12 yr after the diagnosis was evident to them. Although the study neither explicitly conducted longitudinal assessments, nor did include late adolescents in a later age group such as 15-17 yr, which could have provided a better picture of developmental epidemiology.

Standardized psychometric tools were used as far as possible similar to most cross-sectional as well as prospective studies done so far[2,16,27,28]. Only a few studies[23,29] have assessed and shown that intelligence of β-thalassaemic children was within the normal range, as shown in our study (mean IQ of our β-thalassaemic children 107). Neurotic traits, behavioural problems and developmental delays were hardly touched in this subgroup of patients in earlier studies[23,29].

As the mean age and life expectancy of β-thalassaemia major patients has expanded with the advances in medical treatment, psychosocial issues related to quality of life remained an increasingly important focus of attention to assess if the former has brought any paradigmal shift in later. In this study, the quality of life in children was not significantly associated with any of the variables like age of their diagnosis, their Mean IQ, parental psychopathology or overall quality of life of their parents. On the contrary, there was minimal difference in all the variables of quality of life between groups B and C of children. This could be an atypical finding of our study because all such factors have been noted to show their impact in varied degree in most childhood fatal chronic diseases[27,30].

Earlier studies about the psychiatric morbidity in the children of β-thalassaemia major have shown conflicting and curvilinear relationship[17,19,25,26]. In our study we found the cross-sectional prevalence of 27 per cent, which was much less than prevalence of psychiatric diagnosis noted in other studies[17,19,25,26]. Conversely, psychiatric morbidity in group B parents was slightly more than group C parents. Like children, adults vary in their developmental stages and are not homogeneous in their ability to cope. Many adults have limited abilities to abstract and to understand illness. The paradox might also suggest that parents of adolescent thalassaemic children grew up stronger than their pre-adolescent counterpart to keep maintain the mastery and control over the fears of anticipatory death. Overall picture again fits in the developmental epidemiological frame explaining how our study represents the integrated family coalition and likely longitudinal course of psychosocial development, which could not be shown by a mere random, cross-sectional analysis routinely done.

To reach into the depth of emotions about the virtual reactions of the parents immediately after the diagnosis, we developed a reaction to illness questionnaire in Hindi. Although it yet not fully standardized, it has got cognitive, emotional, behavioural spiritual and social domains with 50 items scored from 0-4 on likert scale. This has enabled us to provide succinct picture holding the developmental epidemiological framework as

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subsequently all three groups of parents were analyzed. Reaction toward illness distress was clearly more expressed by parents (group A) who were immediately interviewed rather than the parents in B and C groups, who were asked to open up with their reactions. It was apparent that as the illness progresses in its due course, parents decrease their denial, anger outbursts and internalization as well. Over a period of time using this design, it was seen that most of guilt and self-criticism in the parents (P2 and P3) fades away; rather many of them felt that illness in their child was an opportunity to serve their child. Conversely, their spiritual faith was comparatively loosened in the progressive course as impending death span decreases. This should create enigmatic curiosity in clinicians and researchers about handling the near death children with certain degree of understanding that getting separated from their child surpasses the spiritual faith in most parents and they may need supportive psychotherapeutic mode to provide an auxiliary ego.

There is abundant literature about family coping mechanism in chronic illnesses[24,26,28,30] including thalassaemia major; however, any correlates of maladaptive coping and depressive ideations not mentioned so as to know, whether such strategies secondarily affect their adherence to treatment in later course of time.

This study has several limitations. Despite the accelerated longitudinal design, we could not actually carry out the repeated assessments of each subject of three separate age spanned groups of β-thalassaemia major families, which otherwise would have provided a virtual and real picture of developmental effects rather than hypothetically proceeded period effects. The study did not include late adolescents in a later age group (15-17 yr) which could have provided an expanded and better picture of developmental epidemiology. Poor validity of Reaction to Illness Questionnaire and non usage of the structured diagnostic instrument to make ICD-10 diagnoses in parents were also the limitations.

To conclude, this study expands the literature on psychosocial aspects of β-thalassaemia in several important ways. This study illustrates the developmental epidemiological perspective in β-thalassaemic children and their family using an accelerated longitudinal design. Such design can test the hypothesized etiological or developmental function of a targeted risk factor within a developmental path and may be used in studying the psychological impact of even other chronic illnesses over the life span of an individual for conceptual and holistic understanding.

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