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A Rating Scale for the Functional Assessment of Patients with Familial Dysautonomia (Riley Day Syndrome)

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Abstract

Objective—To develop a reliable rating scale to assess functional capacity in children with familial dysautonomia, evaluate changes over time and determine whether severity within a particular functional category at a young age affected survival.

Study design—Ten functional categories were retrospectively assessed in 123 patients with familial dysautonomia at age 7 years $\pm$ 6 months. Each of the ten Functional Severity Scale (FuSS) categories (motor development, cognitive ability, psychological status, expressive speech, balance, oral coordination, frequency of dysautonomic crisis, respiratory, cardiovascular and nutritional status) was scored from 1 (worst or severely affected) to 5 (best or no impairment). Changes over time were analyzed further in 22 of the 123 patients who were also available at ages 17 and 27 years.

Results—Severely impaired cardiovascular function and high frequency of dysautonomic crisis negatively affected survival ($p < 0.005$ and $p < 0.001$, respectively). In the 22 individuals followed up to age 27 years, psychological status significantly worsened ($p = 0.01$), and expressive speech improved ($p = 0.045$). From age 17 to 27 years, balance worsened markedly ($p = 0.048$).

Conclusion—The FuSS scale is a reliable tool to measure functional capacity in patients with familial dysautonomia. The scale may prove useful in providing prognosis and as a complementary endpoint in clinical trials.

Keywords

disease progression; functional health status; scoring system; survival rate

Familial dysautonomia, also known as Riley Day syndrome and hereditary sensory autonomic neuropathy type III (HSAN III), is a genetic disorder that affects neuronal development and survival of specific neuronal populations resulting in widespread sensory and autonomic dysfunction.¹ The disorder, first described in 1949,² has high morbidity and

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mortality, but as a result of improved supportive treatments prognosis is improving as reflected by improved survival.³ Thus there is increasing concern about quality of life and functional capacity,⁴ and a need to identify which areas of disability have the greatest impact on survival.

Familial dysautonomia is primarily a neurological disorder, but multiple organ systems are affected secondarily resulting in variable expression and myriad clinical symptoms.¹ Pervasive dysfunction of the central and peripheral autonomic nervous systems results in oropharyngeal incoordination with frequent episodes of aspiration pneumonia, esophageal dysmotility and gastroesophageal reflux, blood pressure lability and episodic hypertension, tachycardia and vomiting referred to as “dysautonomic crises”.^{1,5,6} Patients have hypotonia, gait ataxia and, perhaps as a result of sensory impairments, many experience significant distortion of body image, as well as high anxiety levels and learning problems.^{7,8} Kyphoscoliosis, short stature, and poor coordination impose further physical limitations.⁹

Dedicated functional scales have been developed for other chronic illnesses, such as cystic fibrosis,^{10,11} and have been found useful in assessing clinical outcomes. None of the scoring systems, however, are appropriate for the patient with familial dysautonomia because they do not fully encompass all its clinical manifestations.

Thus, we developed a new scale for patients with familial dysautonomia that assessed clinical function in ten different categories. Our aim was to determine if particular areas of function changed over time and whether severity within a particular category at an early age impacted on survival. We anticipated that such an analysis might provide measures that would influence current and future care and yield outcomes that could be used in the design of future treatment trials.

METHODS

A retrospective study was performed using data from patients recruited from the Dysautonomia Center at New York University Langone Medical Center where most patients are seen on an annual basis, which allows for sequential retrieval of data. All participants have consented for participation in a registry as approved by the Institutional Review Board. For the initial functional assessment, age 7 years $\pm$ 6 months was selected because we anticipated that seven-year-olds would have reached milestones appropriate for a school aged child. Using 10-year intervals, two other ages, 17 years $\pm$ 6 months and 27 years $\pm$ 6 months were selected for functional assessment to provide longitudinal information on post-adolescence and adulthood.

One hundred and thirty patients with familial dysautonomia, with typical clinical features and diagnosis confirmed by genetic testing that documented mutations in the *IKBKAP* (I- κ -B kinase complex associated protein) gene, were identified who had reached entry age between January 1975 and April 1, 2007. Complete data were available on 123 (60 female: 63 male). Twenty-two individuals (13 female: 9 male) had data available at all three assessment ages (Figure 1). Reasons for not being included in analysis included death, incomplete data or not reaching the next designated age of assessment.

The Functional Severity Scale

The FuSS is comprised of 10 functional categories. Each category is scored from 1 (worst or severely affected) to a maximum of 5 (best or no impairment).

Motor development—This category scores the age when independent ambulation was achieved, a variable that might impact on survival. The following scoring system was used

and included in survival analyses: (1) patients were unable to ambulate by age 5 years; (2) independent ambulation occurred after 36 months but by 5 years; (3) ambulation occurred from 25 to 36 months; (4) independent ambulation occurred from 18 to 24 months; and (5) independent ambulation was achieved before 18 months, which is within normal developmental limits.

Cognitive ability—This category used the Full scale IQ performance when available, as well as academic performance, and need for special education; for the adult patient, employment capability was utilized: (1) full scale IQ <70 or patients were noneducable; (2) full scale IQ <70 or patients were educable, but academic performance was limited to simple functions and clearly below age level; (3) full scale IQ scores were 70 to 85 and special education class or special school were required; (4) full scale IQ scores were 70 to 85 and patients were able to be mainstreamed for some classes but required resources; and (5) full scale IQ >85 and patients were performing at grade level.

Psychological status—For this category degree of anxiety, emotional maturity, and ability to do self-care were considered: (1) patients were pervasively delayed and independent function was impossible; (2) patients had severe anxiety, phobias, or compulsions and had limited independent function and limited peer relationships (usually need assistance with daily living skills); (3) patients had moderate anxiety and emotional immaturity, but had some peer interaction and were able to do self-care to some extent; (4) patients had mild anxiety and actions that were considered inappropriate for age, but had good peer relationships and were able to do self-care; and (5) patients were emotionally stable and had actions appropriate for age.

Expressive speech—Capability in this category included ability to verbalize and intelligibility of speech: (1) patient had no expressive speech or speech was limited to a few words and phrases; (2) patient had severe dysarthria that was less than 50% intelligible; (3) speech was more than 50% intelligible but there was hyper-resonance with abnormal intonation; (4) speech was completely intelligible but there was mild slurring; and (5) speech was normal.

Balance—Capability in this category was assessed by ability to perform various maneuvers at the time of neurological examination: (1) severe ataxia that prevented independent ambulation and necessitated use of a wheelchair or scooter for mobility; (2) ataxic gait but able to walk with assistance, e.g. use of a walker, cane or someone's arm; (3) can walk a corridor without a walker but gait is very unsteady. Individual cannot hop or balance on one foot; (4) can walk a corridor without a walker but gait may be wide-based; cannot balance on one foot but may be able to hop at least one time on either foot; and (5) can walk a corridor without support and can balance on at least one foot and can hop.

Oral coordination—Capability in this area was based on how the patient derived his or her nutrition (ie, to what degree it was by the oral or enteral route): (1) there was total reliance on gastrostomy for all fluids and calories; (2) all fluid and most nutrition were given via gastrostomy, but some calories were given orally (less than one-third of daily intake); (3) all fluid but minimal calories were via gastrostomy; (4) most of the fluid and calories were via the oral route with only occasional fluid gastrostomy boluses; and (5) all nutrition and fluid were via the oral route

Frequency of dysautonomic crises—To score the frequency of crisis we used a scale that was utilized in two earlier studies:^{12,13} (1) patient experienced almost constant nausea with retching multiple times a day; (2) nausea and retching occurred daily to weekly; (3)

crises occurred six times per year or greater; (4) crises occurred one to five times a year and were usually triggered by intercurrent illness; and (5) patient had not experienced any crises in the year prior to assessment.

Respiratory status—Scores in this category were based upon frequency of pneumonia, oxygen saturation at time of examination, and need for respiratory support during sleep: (1) patients had daytime saturations <92% on room air and a history of more than two lower respiratory infections in the past year, or patients required tracheotomy or ventilation during sleep; (2) patients had saturations equal to or greater than 92% on room air and a history of more than two lower respiratory infections in the past year, or they needed oxygen support during sleep; (3) patients had saturations equal to or greater than 95% on room air and a history of more than two lower respiratory infections in the past year, or they needed supplemental oxygen during sleep only when ill; (4) patients had saturations equal to or greater than 98% on room air and a history of less than two lower respiratory infections in the past year; and (5) patients had saturations equal to or greater than 98% on room air and had not experienced a lower respiratory infection in the past year.

Cardiovascular status—Scoring in this category was based upon reported symptoms: (1) patients had overt syncope (ie, loss of consciousness, with or without documented bradyarrhythmia or asystole); (2) patients had severe orthostatic intolerance with activities that reduce venous return to the heart, such as defecation; (3) patients had presyncopal symptoms, including dizziness, lightheadedness, blurring of vision, or needing to squat periodically but never losing consciousness; (4) patients reported tiredness when standing and walking or complained of leg pain with walking, but did not experience syncope; and (5) patients did not report symptoms associated with orthostatic intolerance.

Nutritional status—Scores in this category were based upon BMI and height: (1) malnourished range was defined as BMI below the 5th percentile no matter what the height attained; (2) patients who were thin and short (BMI was 5th to less than 25th percentiles and height less than the 5th percentile); (3) patients who were short but well nourished (BMI was equal or greater than the 25th percentile and the height was less than 5th percentile); (4) patients who were thin but within the norm for stature (BMI was 5th to less than 25th percentiles and the height equal or greater than the 5th percentile); and (5) patients who were well nourished and of normal height (BMI equal or greater than 25th percentile and height equal or greater than the 5th percentile).

Assessing reliability of the FuSS

Three assessors (FBA, GGVS and DB) independently rated the 123 patients using information from the database. All three raters were familiar with the clinical problems of familial dysautonomia and knew the patients well. Their ratings were used to study inter-rater reliability.

Statistical Analyses

Inter-rater reliability was assessed for each of the functional scores at each time period using intraclass correlation coefficients (ICCs). The ICCs were estimated using two-way mixed effects models in which rater effects were fixed and subject effects were random. The estimated ICCs represented the reliability of average ratings produced by the three raters. Log rank (Mantel-Cox) chi square statistics with four degrees of freedom were used to compare survival among the original 123 individuals to test whether a functional score at 7 years $\pm$ 6 months was associated with survival. In these analyses, patients were grouped and compared by the grade of their 7-year functional score. Kaplan Meier survival curves provided visual comparisons.

Friedman two-way analysis of variance, a nonparametric alternative to repeated measures ANOVA, was used to test for intra-patient differences among measurements using data from the 22 individuals with functional scores measured at ages 7, 17 and 27 years. Separate analyses were performed for each functional category. The test statistic for these analyses was the Friedman chi-square statistic with two degrees of freedom. When differences were found among measurement times, pairwise intra-patient comparisons between time periods were performed using Wilcoxon nonparametric signed rank test, a Z statistic consisting of the standardized sum of ranks with statistical results based on its asymptotic significance level. The Bonferroni correction was used to adjust for the two pairwise comparisons of adjacent time periods.

Nonparametric Mann-Whitney U tests were used to compare all functional scores at age 7 between the 22 patients who lived to age 27 and the 36 patients who died prior to age 27. The test statistic used was a normal approximation to the Mann-Whitney U statistic with statistical results based on its asymptotic significance level. Histograms were drawn for visual comparison of the distributions of scores of the two groups within each functional category.

Statistical analyses were performed using SPSS v16 (SPSS Inc, Chicago, IL). Two-tailed tests were used in all analyses at an alpha level of 0.05.

RESULTS

Inter-rater reliability was excellent for scores in almost all functional categories at all time periods (ICC between 0.75 and 1.0).¹⁴ The three scores with the lowest ICCs, psychological status at age 7, respiratory status at age 17 and cardiovascular status at age 27 were still very good (ICC between 0.69 and 0.74). Not unexpectedly, the best coherence was seen when there were strict objective parameters so that personal judgment was excluded, such as with nutritional status where rating was defined by placement on a percentile chart (ICC=1.0).

Progression of disease

Based on the data from the 22 subjects who survived to age 27, significant changes occurred over time for two functional categories, psychological status ($p=0.01$) and expressive speech ($p=0.045$). The Table displays numerically the progression of disease for the various functional categories by showing the frequency of severity scale values at each age. It can be seen that psychological status scores significantly declined with the transition to adolescence as the percents of patients with better scores (4 and 5) decreased. Expressive speech was the only category that showed significant improvement over time and the percent of best score (5) progressively increased in the adolescent and adult. In the balance category, although there was no overall trend ($p=0.12$), there was significant worsening from age 17 to age 27 years ($p=0.048$).

Impact of severity of a functional category at age 7 years $\pm$ 6 months upon survival

The severity of scores was associated with survival in two of the ten functional category scores, frequency of crisis and cardiovascular status ($p<0.0001$ and $p=0.005$) respectively. In both categories, there were noticeably divergent survival patterns, especially between patients with a score of 2 as compared with the other patients (Figure 2). Patients with severe scores of 2 died at earlier ages than the other patients. In both functional categories there were only one or two patients with the most severe score of 1.

Comparison of functional scores for patients who survived to age 27 and those who did not

When scores were compared between the 22 patients who survived to age 27 years and the 36 patients who died prior to 27 years (Figure 3), scores for six functional categories were significantly higher for the patients who survived to age 27 (cognitive ability, $p=0.021$; psychological status, $p=0.028$; expressive speech, $p=0.009$; balance, $p=0.003$; oral coordination, $p=0.045$; frequency of crisis, $p=0.011$). In these six categories, the frequency of grades 4 and 5 (the better scores) were notably increased for the patients who reached age 27.

DISCUSSION

Familial dysautonomia is a devastating genetic disorder with a complex neurological phenotype that severely affects sensory function, balance, swallowing, esophageal and gastrointestinal motility and cardiovascular control.^{1,5,6} The disorder is caused by mutations in the *IKBKAP* (I- κ -B kinase complex associated protein) gene on chromosome 9q. Although >99% of patients with familial dysautonomia are homozygous for the IVS20+6T>C mutation,¹⁵ disease expression is extremely variable. It is believed that the disorder worsens over time,^{1,16} but available survival statistics do not take into account how phenotypic variability may affect function, how function may change over time, and which areas of disability may impact on survival.

The FuSS score is the first attempt to develop a functional capacity scale for patients with familial dysautonomia to help chart the natural history of the disorder, as well as to study its impact on quality of life. We believe this scale describes functional areas that are unique to the patient with familial dysautonomia. The items for the tool were selected based upon clinical experience treating patients with familial dysautonomia at a single center. The ten categories were identified as those of utmost concern to treating physicians and families. We developed our scales to be as objective as possible. Although the three raters used in our study were familiar with the subjects included in this study, every item used to score a functional category could be found in a subject's medical records. For this reason, we are confident that the high degree of reliability will be similar in future studies.

One major aim of this study was to follow changes in function over time. Thus we developed our scores using five item Likert scales because these scales enabled us to adequately define gradations of the functional categories. Using the FuSS we were able to demonstrate that scores obtained at 7 years $\pm$ 6 months for two of the ten functional categories, frequency of crisis and cardiovascular status, were significantly associated with survival. (Figure 2). The worse survival noted for patients with very frequent crises may relate to their need for frequent medication or inability to maintain adequate nutrition or more labile autonomic function. Survival results for cardiovascular status demonstrate that the group at highest risk contained patients with a score of 2, who were those patients with most severe orthostatic intolerance and frequent syncope triggered by minor hemodynamic stresses, such as defecation (Figure 2, B). These patients may be most susceptible to the consequences of afferent baroreceptor failure.⁶ Interestingly an early marker of neurological development, the age at which a child with familial dysautonomia attains independent ambulation (ie, motor development scores), did not affect survival. The survival analyses that we performed on our particular dataset show that some contiguous categories show similar survival patterns. However we consider it premature to combine categories based on these initial results. If the patterns that we see persist in analyses of other datasets, we will consider combining contiguous categories in the future.

The FuSS scores also provide insight into the natural history of familial dysautonomia. The population of 22 who survived to age 27 years was notably different from the population of 36 who died prior to age 27 years (Figure 3). The population of 22 who survived, had better scores in six functional categories, cognitive ability, psychological status, expressive speech, balance, oral coordination, and frequency of crisis suggesting that these surviving patients perhaps had milder disease expression. For example, better expressive speech and better oral coordination may reflect better brainstem function or relate to an individual's ability to better protect his/her airway and respiratory tract. Further analysis of the 22 individuals who had survived to age 27, showed that expressive speech improved significantly over time, which may reflect the impact of continued therapy and social stimulation. However, even in this relatively milder group of survivors, psychological status significantly declined over time ($p=0.01$), especially between ages 7 and 17 years (ie, the transition to adolescence) (Table). This decline in psychological status scores across the adolescent years is consistent with previous reports of emotional immaturity and anxiety in patients with familial dysautonomia.⁸ Balance was another functional category that was affected by time; worsening balance was most apparent by early adulthood as the combined percentage of better scores (ratings of 4 and 5) at ages 7 and 17 was 59 % but then decreased to 36 % at age 27.

The study design is limited by the fact that the raters were familiar with some of the patients whose records they analyzed, specifically those patients that were still surviving in the last 10 years of the analysis. Another possible limitation of our functional severity scale is that it does not include spine curvature, vision, or renal function, which are also important factors for quality of life and independence. However at the initial examination age of 7 years, these issues are not usually apparent and we wished to focus on factors that would appear early and provide families and caregivers with some prognostic information. For example, parents and caregivers now could be reassured that traditional markers such as age of walking does not correlate with cognition or overall function but that better control of crisis frequency and blood pressure variability could significantly affect outcome. Furthermore, now that it is known that familial dysautonomia is caused by a tissue-specific splicing mutation in the *IKBKAP* gene, various modifiers of gene expression have been proposed as therapeutic agents.^{17,18} None have yet been submitted to long-term trials. However, as such trials are initiated the FuSS could prove valuable as an outcome measurement tool to determine if the natural history of the disorder is modified.

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ABBREVIATIONS

FuSS	Functional Severity Scale
ICC	intraclass correlation coefficient

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Figure 1.
Description of the total study population.

Figure 2.

Kaplan Meier survival plots of patients with familial dysautonomia grouped by scores. A, Frequency of crisis at age 7 years. B, Cardiovascular status at age 7. Green line, patients with scores of 2 (n=24 for crisis; n=13 for cardiovascular status); blue line, patients with score of 3 (n=15 for crisis; n=20 for cardiovascular status); purple line, patients with score of 4 (n= 44 for crisis; n=40 for cardiovascular status); orange line, patients with score of 5 (n= 38 for crisis; n=49 for cardiovascular status). The severity score of 1 is not represented because A, only two patients had this score, and B, only one individual had this score.

Figure 3.

Distributions of scores within specific functional categories at age 7 years. Black bars, percentage of patients with familial dysautonomia who survived to age 27 years (n=22); grey bars, percentage of patients with familial dysautonomia who died prior to age 27 years (n=36).

Table 1

Percent of Scores within Specific Functional Categories: Comparison of Changes in Scores from 7 years to 17 years to 27 years (n=22).

Score	Motor Development			Cognition			Psychological Status			Expressive Speech			Balance		
	Age 7	Age 17	Age 27	Age 7	Age 17	Age 27	Age 7	Age 17	Age 27	Age 7	Age 17	Age 27	Age 7	Age 17	Age 27
1	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
2	9.1	9.1	9.1	0	0	9.1	0	18.2	18.2	0	0	0	0	4.5	22.7
3	27.3	27.3	27.3	18.2	18.2	9.1	4.5	13.6	18.2	4.5	4.5	9.1	40.9	36.4	40.9
4	54.5	54.5	54.5	22.7	27.3	36.4	68.2	54.5	45.5	54.5	31.8	18.2	50.0	31.8	18.2
5	9.1	9.1	9.1	59.1	54.5	45.5	27.3	13.6	18.2	40.9	63.6	72.7	9.1	27.3	18.2
Score	Oral Coordination			Crisis Frequency			Respiratory Status			Cardiovascular Status			Nutrition		
	Age 7	Age 17	Age 27	Age 7	Age 17	Age 27	Age 7	Age 17	Age 27	Age 7	Age 17	Age 27	Age 7	Age 17	Age 27
1	0	4.5	0	0	0	0	0	0	0	0	0	4.5	18.2	27.3	27.3
2	0	0	4.5	4.5	13.6	4.5	0	0	4.5	4.5	18.2	9.1	18.2	4.5	4.5
3	27.3	22.7	22.7	13.6	13.6	18.2	36.4	50.0	31.8	22.7	31.8	18.2	31.8	50.0	50.0
4	0	4.5	9.1	40.9	40.9	45.5	45.5	18.2	22.7	45.5	40.9	63.6	4.5	9.1	4.5
5	72.7	68.2	63.6	40.9	31.8	31.8	18.2	31.8	40.9	27.3	9.1	4.5	27.3	9.1	13.6