Anorectal malformations and Hirschsprung disease
Functional and psychosocial challenges
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Citation for published version (APA):

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Persistent parental stress in parents caring for a young child with an Anorectal Malformation or Hirschsprung disease

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Submitted
Abstract

Objectives
Anorectal malformations and Hirschsprung disease are chronic congenital disorders, which can be very stressful for parents. The objective of this study was to investigate parental stress, caused by the diagnosis and the treatment of their child with an anorectal malformation or Hirschsprung disease, and to assess whether parental stress changes over time.

Design
Since 2012 we started the KLANKbord-study: a multicentre, longitudinal QoL study for recently diagnosed patients with an anorectal malformation or Hirschsprung disease and their parents.

Methods
To measure parental stress we used the Pediatric Inventory for Parents (PIP). Both the frequency (PIP-F) of stressful illness-related events and the difficulty (PIP-D) parents experience with these events were measured. Three measurements were done: 6-12 months, 2 years and 3 years after the diagnosis.

Results
In total parents of 25 patients were eligible for this study (8 anorectal malformations and 17 Hirschsprung disease). During measurement one the mean PIP-F score was 82.11 (SD 23.50) and the PIP-D score was 73.71 (SD 23.88). PIP-F score for anorectal malformation was 92.13 (SD 25.05) and PIP-D score was 79.71 (SD 33.63). PIP-F score for Hirschsprung disease was 77.39 (SD 21.90) and PIP-D score was 69.06 (SD 19.53). No statistically significant differences were found between all measurements (total group and anorectal malformations or Hirschsprung disease individually).

Conclusions
The PIP scores indicate that from half a year up to a year after diagnosis, parents experience levels of stress comparable to parents of patients with other chronic diseases. The stress level remains elevated up to three years after diagnosis. This means that interventions for parental stress reduction can be considered, even years after the diagnosis.
Introduction

Anorectal malformations and Hirschsprung disease are both congenital disorders, usually diagnosed soon after birth. These disorders can have an enormous influence, both on the child and his or her parents because of the consequences and chronic character these diseases have. The incidence of anorectal malformations and Hirschsprung disease is about 1 in 5000 live births, resulting in a total of approximately 80 children born in the Netherlands each year, with either an anorectal malformation or Hirschsprung disease.\(^1\) Anorectal malformations comprise a wide spectrum of anomalies of the anus and rectum, and 43% to 71% of the patients also have additional congenital anomalies, often involving the urogenital tract.\(^2\) The malformation can be as little as an anal membrane, but it can also involve rectourethral, rectovesical, rectovestibular fistulae or even a cloacal malformation.

In patients with Hirschsprung disease there is a variable length of aganglionic, non-peristaltic colon. Meaning this part of the colon is unable to relax leading to severe constipation.\(^3,4\) Anorectal malformations are often diagnosed directly after birth, however Hirschsprung disease can be a difficult diagnosis to make. Patients sometimes have to undergo numerous investigations or interventions before the diagnosis is definitive. This can be a stressful period for the parents. Almost all of the patients with an anorectal malformation or Hirschsprung disease are surgically corrected shortly after the diagnosis is made, but in a number of patients a (temporary) ostomy needs to be created before the definitive surgical correction.

The operation is an essential part of the treatment but not a definite cure. It may be the start of a chronic course with many disease related sequelae.\(^5\) Patients with an anorectal malformation or Hirschsprung disease may, for instance, experience mild to severe defecation disorders and other disease-specific problems throughout their entire life.\(^6,7\) And sometimes it is necessary for parents to perform invasive procedures with their child, such as anal dilatations or rectal wash outs. All of this not only affects the child’s QoL but also puts high pressure on the parents ability to cope with all the stress associated with dealing with a child with a chronic disease.\(^8,9\)

In a number of other chronic diseases an elevated level of parental stress has proven to have a negative influence on the child’s disease.\(^10-14\) High levels of parental stress are associated with a delay in the recovery of a child after a traumatizing event, for example a cancer diagnosis.\(^15\) Early intervention focusing on the parental stress, in a family system approach, has had a positive influence on both parents and the child. This effect can be measured until late childhood. The study from Landsem et al. showed only a glimpse of
what could be accomplished if more was known about parental stress and its interventions, which can lead to a reduction of stress. In patients with Hirschsprung disease so far no studies have been performed investigating parental stress. For patients with an anorectal malformation 3 studies have been done on the subject of parental stress related to the disease and its treatment. All these studies were based on interviews, leaving room for interpretation and no options for statistical analysis. To obtain a better understanding of parental stress related to an anorectal malformation or Hirschsprung disease in their offspring a multi-dimensional assessment, specific to the circumstances of parents of these children is needed. In addition more information about the way this parental stress develops and changes over time must be obtained in order to address the need for and consequently the development of early family focussed interventions.

The aim of this present study is to investigate the level of parental stress of parents caring for a young child with an anorectal malformation or Hirschsprung disease. And to assess whether this parental stress changes in time. Our hypothesis is that stress will be reduced over time, because parents learn to cope with the disease of their child.

Material and Methods

Patients
In 2012 we started the KLANKbord study in the Netherlands. This is a multicentre, longitudinal QoL study of patients with an anorectal malformation or Hirschsprung disease and their parents. In this particular study we included patients that were born from 2012 until present date. Parents of the patients are approached within several weeks after the diagnosis is made, and invited to participate in this study. They receive a set of questionnaires every 6 months until their child reaches the age of 4 years. From the age of 4 years old they will receive questionnaires once a year until the age of 17. If parents do not want to participate, or do not have the ability to speak Dutch or English, they are not included.

Ethical approval
Since this study focuses on evaluation of health care practice, specific approval of the medical ethical board was waived. Informed consent was given by all parents.

Questionnaire
The Pediatric Inventory for Patents (PIP) is a questionnaire completed by parents together, starting 6 months after the diagnosis anorectal malformation or Hirschsprung disease was made. The PIP, designed by Streissand et al., investigates areas of anxiety and concern experienced by parents caring for a child, with an illness, without being
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limited to a specific one. It combines disease related measures with generic measures so that these dimensions can be investigated in a quantitative fashion. One of the assets of the PIP is that parents are asked to rate both the frequency (PIP-F) of stressful illness-related events and the difficulty (PIP-D) they experience with these events since this gives a better insight on both coping and stressors. All questions are answered for 42 disease related events in a 5-point scaling for each subscale (1 = “not at all”, 5 = “extremely”). The 42 questions are divided amongst four domains: communication with the family/medical professional (e.g. “talking with a doctor”), emotional functioning (e.g. “trying not to think about my family’s difficulties”), medical care (e.g. “making decisions about medical care”), role function (e.g. “trying to attend to the needs of other family members”). For every completed form the frequency and difficulty scores are generated for each domain and as an overall score (maximum score 210). Higher scores indicate higher levels of stress. The PIP does not have a cut of score from which the label “stress” can be applied. Instead it is useful for comparison between different diseases or, as in this study, within the same patient group at different moments in time.

The PIP has been widely validated in numerous diseases such as cancer, diabetes and inflammatory bowel disease (IBD). It has been translated in multiple languages, including the Dutch language, which is validated and considered satisfactory.

Measurements

This study comprises 3 measurements. Measurement 1 was 6 to 12 months after the diagnosis. Measurement 2 was two years after the diagnosis and measurement 3 three years. To observe if stress changed over time a baseline PIP score for comparison was necessary. That is why the main criterion to be included in this study was the availability of the baseline score at measurement one.

Missing data

During measurement 1, three out of 25 patients had missing data. One of them missed 1 answer out of 84 and two missed 4 out of 84. We used the row mean to fill in for these missing data. During measurement 2 we only filled in the missing data (using feed forward) if there were 6 items or less missing. This was the case for 4 patients. A feedforward system was used, using the measurement of moment 1 as a substitute. The same was done for measurement 3, for which moment 2 was used as a substitute.

Demographic analysis

The analysis for differences between moment 1 and 3 was based on gender, having more children, child’s position compared to possible brothers/sisters, parental education level and family income. These factors were chosen as confounding factors, because of earlier studies suggesting their influence on parental stress.
**Statistical analysis**

Descriptive statistics were used to search for missing data in the three different measurements. To review if the parents were equally distributed, during different measurements, a Chi-squared test was performed for a number of socio-demographic factors known to have an influence on parental stress (such as parental level of education). With all the data complete we calculated the PIP-D and PIP-F scores. We screened for a normal distribution by looking at the histogram of the data. A paired-T test was done on these scores to measure the significance of differences in scores. Also, a paired-T test was done on anorectal malformation and Hirschsprung disease cases separately. Level of statistical significance was set at \( p < 0.05 \).

Delta’s scores, the difference between one measurement and the next moment of measuring, were also computed. This was done to observe whether the mean difference was influenced by extremely high and low PIP scores or if PIP scores were centred closely around the mean.

All analyses were performed with the Statistical Package for Social Sciences (SPSS version 23).

**Results**

**Patients characteristics**

A total of 61 patients were enrolled in the KLANKbord study at the time of this analysis. One of the main criteria was the availability of the baseline list (6-12 months after the diagnosis). Thirty-four patients dropped out because of different reasons (Figure 1). Two patients were included in the KLANKbord study at older age. Leaving 25 patients eligible for this study. Anorectal malformations were diagnosed in 8 cases of which 5 males and 3 females. Hirschsprung disease was diagnosed in 17 cases of which 12 males and 5 females. During measurement 2, the questionnaires of 21 patients were available. Two patients did not yet reach the age of two, 2 patients terminated their enrolment in the study. During measurement 3, 11 patients were included. Nine patients did not yet reach the correct age and 1 failed to complete the PIP questionnaire (Figure 2).

**PIP scores analysis**

The histogram of the data showed a normal distribution. PIP scores are presented in Table 1. During measurement one the mean PIP-F score was 82.11 (SD 23.50) and the PIP-D score was 73.71 (SD 23.88). We also compared anorectal malformations and Hirschsprung disease individually. PIP-F score for anorectal malformations was 92.13 (SD 25.05) and PIP-D score was 81.65 (SD 31.65). PIP-F score for Hirschsprung disease was 77.39 (SD 21.90) and PIP-D score was 69.98 (SD 19.25). An independent sample T-test
showed no significant difference between the separate anorectal malformations and Hirschsprung disease PIP-F ($p = 0.385$) and PIP-D ($p = 0.067$) scores.

All the Chi squared analyses on demographic differences between moment 1 and 3 scored a $P > 0.1$ suggesting no differences between these groups. No statistically significant differences were found between all moments suggesting no change in PIP scores over the course of three years after diagnosis (Table 2). This was also the case when a paired T-test was performed on all moments for anorectal malformations and Hirschsprung disease individually.

Using the delta’s scores for PIP-D and PIP-F 6 histograms and 2 boxplots (Figure 3 and Figure 4) were created to observe the variance of the dataset. These histograms show that, for both PIP-D and PIP-F scores at all measurements, there are parents who increase or decline in their stress. The difference between the two however is minor; most deltas for every measurement were between -20 and +20. There was no trend in the deltas.
outside this range other than that most of these scores were considered outliers in the boxplot. They were divided equally between above and below the range.

<table>
<thead>
<tr>
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</tr>
</thead>
<tbody>
<tr>
<td>PIP-F Total (SD)</td>
<td>82.11 (± 23.50)</td>
<td>77.40 (± 23.01)</td>
<td>75.09 (± 15.73)</td>
</tr>
<tr>
<td>PIP-D Total (SD)</td>
<td>73.72 (± 23.88)</td>
<td>68.33 (± 20.73)</td>
<td>66.10 (± 29.05)</td>
</tr>
<tr>
<td>PIP-F ARM (SD)</td>
<td>92.13 (± 25.05)</td>
<td>80.00 (± 15.36)</td>
<td>80.00 (± 19.31)</td>
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<tr>
<td>PIP-D ARM (SD)</td>
<td>81.65 (± 31.65)</td>
<td>61.33 (± 12.85)</td>
<td>59.00 (± 11.47)</td>
</tr>
<tr>
<td>PIP-F HD (SD)</td>
<td>77.39 (± 21.90)</td>
<td>76.29 (± 26.05)</td>
<td>71.00 (± 12.33)</td>
</tr>
<tr>
<td>PIP-D HD (SD)</td>
<td>69.98 (± 19.25)</td>
<td>71.13 (± 22.92)</td>
<td>73.20 (± 40.52)</td>
</tr>
</tbody>
</table>

ARM=Anorectal malformation; HD=Hirschsprung disease

Table 1: Total and individual PIP scores

<table>
<thead>
<tr>
<th>Pair</th>
<th>N</th>
<th>T</th>
<th>Significance (2-tailed)</th>
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<tr>
<td>F1 * F2</td>
<td>21</td>
<td>0,404</td>
<td>0,691</td>
</tr>
<tr>
<td>F2 * F3</td>
<td>11</td>
<td>0,122</td>
<td>0,906</td>
</tr>
<tr>
<td>F1 * F3</td>
<td>11</td>
<td>0,705</td>
<td>0,497</td>
</tr>
<tr>
<td>D1 * D2</td>
<td>21</td>
<td>0,552</td>
<td>0,587</td>
</tr>
<tr>
<td>D2 * D3</td>
<td>10</td>
<td>-0,574</td>
<td>0,580</td>
</tr>
<tr>
<td>D1 * D3</td>
<td>10</td>
<td>0,812</td>
<td>0,438</td>
</tr>
</tbody>
</table>

F = PIP-Frequency score; D = PIP-Difficulty score; Fx = Number for x is number of measurement

Discussion

The aim of this present study was twofold: first we wanted to present a baseline PIP-D and PIP-F score for parents caring for a child with an anorectal malformation or Hirschsprung disease in order to contribute to the growing list of known chronic disease-related PIP scores. Secondly, we wanted to investigate the influence of time on parental stress. By knowing the influence of time, it is possible to reason whether or not an intervention should be done and the timing thereof.

Our baseline scores for anorectal malformations and Hirschsprung disease together (PIP-F: 82.1 (SD 23.50), PIP-D: 73.7 (23.88)) are comparable, however slightly lower, to other chronic diseases such as inflammatory bowel disease (IBD) and diabetes.\textsuperscript{11,13} This minor difference could be caused by the age of the children at the time of the study. The patients with IBD had a mean age of 15,4 years when they were diagnosed and the
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patients with diabetes a mean age of 12.9 years. Our patients were diagnosed between the age of one day and 3 months. During puberty, denial of the disease leads to low child self-care behaviour. And this has been shown to cause an increase in parental stress.\textsuperscript{11} Another possibility is that our scores are an underestimation. Out of the 25 couples in this study, 16 had an academic education. Also 17 out of 25 had a family income of +40,000 euros a year.

\begin{figure}[ht]
\centering
\includegraphics[width=\textwidth]{delta_scores_F.png}
\caption{Boxplot Delta scores PIP-F \newline Delta 1F = PIP-F2 – PIP-F1 (N=20); Delta 2F = PIP-F3 – PIP-F2 (N=10); Delta 3F = PIP-F3 – PIP-F1 (N=11)}
\end{figure}

\begin{figure}[ht]
\centering
\includegraphics[width=\textwidth]{delta_scores_D.png}
\caption{Boxplot 2 Delta scores PIP-D \newline Delta 1D = PIP-D2 – PIP-D1 (N=21); Delta 2D = PIP-D3 – PIP-D2 (N=10); Delta 3D = PIP-D3 – PIP-D1 (N=10)}
\end{figure}
These are high percentages compared to the general population’s level of education and income. High levels of academic education and income have shown to reduce parental stress.\textsuperscript{7,11} Another reason for a slightly lower score could be the fact that the children were still young and not potty trained yet. Fecal continence is not yet an issue for these parents. As a toddler it is completely normal to wear a diaper and not to be fully continent for feces, but when these children grow older and go to school this problem could cause more parental stress.

In many studies anorectal malformations and Hirschsprung disease are combined, because patients with either disease can suffer from the same functional problems during follow-up.\textsuperscript{23-26} However children with an anorectal malformation often have multiple anomalies involving the urogenital tract in addition to anomalies in the gastro-intestinal tract, whilst patients with Hirschsprung disease have a disease more often limited to the gastro-intestinal tract only. This difference in disease complexity could lead to more operations and also involvement of multiple specialities. All of which could lead to elevated stress. This also might be the reason for the seemingly difference in parental stress of parents of children with an anorectal malformation compared to Hirschsprung disease, although not significant. It is possible that it would be a significant difference if more children with an anorectal malformation had been included in this study.

The second aim was to investigate the influence of time on parental stress. Our study suggests that there is no significant change in the level of stress up to three years after the diagnosis. However a non-significant decline in stress is seen, meaning that the stress levels possibly could be significantly lower in future follow-up.

By examining the deltas through both histograms and boxplots, we showed that our mean difference was not caused by extremely high and low PIP scores. This shows that our dataset had a small variance. Mean scores for every delta were close to zero with few outliers. All, but one, of the individuals with a larger than average delta observed, were not consistent over more than one delta score. Only patient number 18 was elevated both in delta2-3D and delta2-3F. This patient needed invasive procedures concerning his disease, which could on its own lead to elevated levels of parental stress. In this group we did not have enough patients in need of invasive procedures to perform a statistical analysis and therefore this is only speculation. More research has to be done on this specific group of patients. By following a broader group over a longer period of time this could be realised.

The other outliers could be caused by a temporarily stress elevating situation (eg. medical treatments, unpleasant results from the hospital) in the vicinity of the moment the parents completed the questionnaire. Over the course of time these elevations show
to be short-lived and all outliers deltas returned closely to mean scores in the following delta. This suggests no consequent elevation or declination of stress.

All stated above suggests that even if stress declines over time, the decrease may take years. It also shows that interventions in order to reduce parental stress, and thereby improving the health situation of the child diagnosed, may be useful even years after the diagnosis. Although stress may be constantly elevated during the first years after diagnosis, we have seen in another study within the same patient population that maternal anxiety diminishes.27 This means that although the anxiety, caused by hearing the diagnosis anorectal malformation or Hirschsprung disease becomes less after a year, it does not reduce the parental stress in years to come. Medical treatments (surgery), hospital visits, bowel management programs and functional problems do not stop after this first year. And this brings us to the conclusion that anorectal malformations and Hirschsprung disease are chronic diseases, which also have chronic consequences on parental stress. At least for the first three years.

One of the important limitations of this study is the relative small number of parents eligible for this analysis. Also the fact that some parents stopped completing our questionnaires is a limitation of this study. However analysing their results, we noticed that the same proportion of parents scored higher versus lower than the mean score, both on PIP-D and PIP-F. The fact that the percentage of parents of patients with an anorectal malformation is relatively low compared to Hirschsprung disease is another limitation.

One of the problems in using the PIP to follow-up on stress over time is that nothing is known about the clinical significance in the change of PIP-scores. The PIP assesses the sum of all stressors and that is why a beneficial but specific intervention on a specific domain (eg. talking to doctors) could be clinically relevant but would not lower the PIP as a whole. Another difficulty is that the PIP does not show why parental stress is elevated in the first place. This is a limitation of this specific questionnaire. Both problems could be solved in the future by also using the Coping Health Inventory for Parents (CHIP). The CHIP is an instrument, which measures parental coping patterns at three subscales: firstly maintaining family integration, second maintaining social support and third understanding healthcare situations. By using the CHIP in the future we could search for group specific coping failures (for example: lower education may lead to bad coping in the subscale of understanding healthcare situations). Interventions aiming to enhance failing coping mechanisms have shown to be effective in stress reduction in chronically ill children and their parents.16,28-30

Subscale specific coping interventions could be developed if scale specific failure was to be found in parents of children with an anorectal malformation or Hirschsprung disease.
This is just one of the few possibilities. More research should be done on possible interventions and their effect on parental stress.

This is the first study to use an illness-related score of parental stress to describe the experienced frequency and difficulty of stressful events by caregivers of anorectal malformation or Hirschsprung disease patients. The PIP scores in this study indicate that from half a year up to a year after diagnosis, parents experience elevated levels of stress, which remain elevated up to three years after diagnosis. This suggests that the elevation of parental stress is not a temporary condition initiated by the diagnosis, but rather a long-term stable state. If this is the case it may increase the need for interventions aiming at stress reduction, and it entails that these interventions may be useful even years after diagnosis. More research has to be done on the origin of parental stress and effective interventions and longer follow-up is needed.
Reference List


