

From the Department of Woman and Child Health
The Unit of Pediatric Surgery
Karolinska Institutet, Stockholm, Sweden

CHILDREN WITH HIGH AND INTERMEDIATE IMPERFORATE ANUS

**Aspects of care and psychosocial effects
of the malformation**

Maria Öjmyr-Joelsson



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To my mother

ABSTRACT

The overall aim of this thesis was to investigate the experiences of children with high and intermediate imperforate anus and of their parents; how they have experienced care and treatment and what consequences the malformation involve for them.

Materials: For *Paper I* three patients with high imperforate anus and their families were interviewed. The patients were born before 1987 and at the time of the interview they were 12, 14 and 22 years old. *Paper II* and *III* are based on the same study sample. Twenty-five children with high or intermediate imperforate anus born 1987-1992 and their parents participated in the index group. Two groups of children similar to each other in age and gender distributions, along with their parents, served as controls.

Methods: In *Paper I* the interviews were conducted at the hospital. The interview guide consisted of open-ended and structured questions, and the sessions covered many different aspects of the child's malformation, as well as treatment and psychosocial consequences of the defect. In *Papers II* and *III* the instruments used to gather data were questionnaires developed on the basis of the previous interview study. The children's questionnaire consisted of 45 items divided into psychosocial domains and one physical domain. The parent's questionnaire consisted of 59 items covering various domains such as the child's hospital care, questions about psychosocial issues concerning both the child and the parent and finally the child's physical functioning associated with the malformation. Child Behavior Checklist was filled in by the parents. Teachers Report Form was filled in by the children's teachers.

Results: In the analysis of the interviews four categories related to effects of the malformation emerged: physical, emotional, social, and family. The physical and emotional effects of the child's malformation influenced the family's experiences. Children with imperforate anus exhibited an overall positive attitude. They reported that they enjoyed school, had good relationships with schoolmates, had several friends, and participated in leisure activities. Constipation occurred more frequently than in control groups. Answers from parents of children with imperforate anus indicated lower values on variables concerning care, information, and involvement in the child's healthcare treatment, compared with the two control groups. No significant differences on these issues were found between the answers of the mothers and fathers of children with imperforate anus. In the CBCL, the children in the index group were assessed to have significantly more emotional/behavioral problems than the children in control groups. In the TRF, the teachers reported few symptoms for the index group children.

Conclusions: Parents of children with imperforate anus had experienced suffering and many difficulties associated with the malformation. Children with imperforate anus in this study did not seem to have any psychosocial impairment, though their parents stated that they experienced leakage, constipation, and fecal odor. The children might have some psychosocial difficulties according to parents but not according to their teachers. Parents of children with imperforate anus experienced less satisfaction with medical care and the support received had been insufficient. An open communication and an interaction between healthcare professionals are important. Psychosocial support for children with imperforate anus and their parents has been limited in our clinic earlier, and a liaison between the pediatric surgery unit and the child and adolescent psychiatry unit would be desirable.

Key-words: imperforate anus, children, fecal incontinence, psychosocial, parental experiences, care.

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LIST OF ABBREVIATIONS

CBCL	Child Behavior Check List
Imp anus	Imperforate anus
JCA	Juvenile Chronic Arthritis
MACE	Malone's Antegrade Continence Enema
PSARP	Posterior Sagittal Anorectoplasty
TRF	Teachers Report Form
VAS	Visual Analogue Scale

1 INTRODUCTION

A child's birth is one of the most important experiences in parents' lives. It is normally associated with feelings of joy, but when the newborn child is ill and needs hospital care the presumed positive birth event is transformed into another dimension (Hall, 2005b; Hopia, Tomlinson, Paavilainen & Astedt-Kurki, 2005). Having a child with imperforate anus raises many questions for the parents, and the care of the child is different in several ways from that of other children. Imperforate anus is an uncommon malformation of the anus and the affected child may have several difficulties at birth that derive from the lesion.

Imperforate anus has been described in literature since ancient times, and various surgical interventions are mentioned as lifesaving measures (Stephens & Smith, 1971). Nowadays surgical methods are advanced: a new anus is constructed, and the improved follow-up treatment makes it possible for a child with imperforate anus to have a fairly normal life. Since constipation and fecal incontinence are common among children with imperforate anus, the parents caring for the child's problems must prevent and reduce the inconveniences in order for the child to be able to live without too many restrictions. Fecal incontinence is shameful, and can cause psychosocial problems throughout the child's growing up, such as being teased and called names in school. Experiences like this could also affect the children and lead to secretiveness and devious behavior (Ludman & Spitz, 1996).

Parents caring for children with disabilities have been studied, but there are few studies on caring for a child with imperforate anus. The psychosocial consequences for children with imperforate anus and their parents, and how the children's difficulties affect these families' lives have only been described in very few studies. We meet these children and their parents in our daily work, as the malformation entails frequent visits to the pediatric surgery unit. We as professionals may have underestimated the care support these families need. Parent groups may be an alternative, to support the parents and their children. This study was aimed at investigating experiences of children with high and intermediate imperforate anus and their parents - how they have experienced care and treatment and what consequences the malformation involves.

2 BACKGROUND

2.1 IMPERFORATE ANUS

Imperforate anus is a malformation of the anorectum. Historically this malformation has been known since antiquity. One of the first mentioned surgeons who dealt with it was Paul of Aegina (A.D. 625-690). He performed an operation to open an orifice into the rectum, then kept it open with dilations (Stephens & Smith, 1971). The surgical technique improved and a second landmark was when Amussat developed the proctoplasty in 1835 (Stephens & Smith, 1971).

2.1.1 Incidence and classification

The incidence of imperforate anus is one in every 4-5000 child born. The malformation is somewhat more frequent among boys than girls. The most common defect among boys with imperforate anus is the rectourethral fistula and the most common defect among girls with imperforate anus is the rectovestibular fistula (Peña, 2000). Classifications of anorectal malformations differ in literature though the severity can be divided in two categories - high and low lesions. In the high lesions, the development of the rectum has closed above the levator muscle. In the low lesions, the development of the rectum has closed below the levator muscle. There is also one type of imperforate anus that can be categorized as intermediate: it does not fit into either of the groups mentioned above. These intermediate malformations are generally treated as high lesions (Kiely, 1998).

An international classification of imperforate anus was proposed in 1970 (Stephens & Smith, 1971) and some years later the Wingspread Conference defined classification, identification, and assessment of surgical treatment of anorectal anomalies (Stephens & Durham Smith, 1986). Experienced surgeons (Kiely, 1998; Peña, 2000) have declared that neither of these classifications are useful in practice, and have proposed a more therapy-orientated classification of the malformation that makes a prognosis for the functional outcome (see below, 2.2 and 2.3).

2.1.2 Associated malformations

It is not unusual to find other malformations in addition to the imperforate anus in 50-60% of the children affected. These associated malformations can include

cardiovascular anomalies, gastrointestinal anomalies, vertebral anomalies, genitourinary defects (Kiely, 1998).

2.2 MALE DEFECTS

2.2.1 Cutaneous fistula

In the low defects the lowest part of the rectum is anteriorly mislocated and placed somewhere along the midline raphe, on the scrotum, or at the base of the penis. The anal fistula is often narrow (Peña, 2000).

2.2.2 Rectourethral fistula

This is the most common defect among male patients. In rectobulbar fistula the fistula is located in the lower part of the urethra. Rectoprostatic fistula is located in the prostatic portion of the urethra. The rectum and the urethra have a common wall above the fistula. A distension of the rectum is common and the most lateral part is surrounded by the levator muscle. The external sphincter muscle complex is located between the rectum and the perineal skin: when contracted it elevates the skin of the anal dimple. Children with low urethral fistulas, rectobulbar fistula, often have well developed muscles, sacrum, midline, and an anal dimple. In children with higher urethral fistulas, rectoprostatic fistula, the risk is higher for poor muscles, sacral anomalies, and a flat perineum with absence of midline groove and anal dimple (Peña, 2000).

2.2.3 Rectovesical fistula

The rectum opens into the bladder neck with a fistula. Among boys with this condition the levator muscle, the external sphincter muscle complex, is generally poorly developed. The sacrum is often deformed. A flat bottom is often seen because of the poor muscle development (Peña, 2000).

2.2.4 Unusual male defects

Anorectal agenesis without a fistula and rectal atresia appear but are unusual defects.

2.3 FEMALE DEFECTS

2.3.1 Cutaneous fistula

The lowest portion of the rectum is located just in front of the normal place for the anus though the rest of the rectum has developed all the way through the muscle complex and is located within the sphincter mechanism. Rectum and vagina are well separated (Kiely, 1998).

2.3.2 Rectovestibular fistula

The rectum opens in the vestibule of the female just behind the hymen. The vagina and the rectum are separated by a thin wall. These girls usually have well-developed muscles and a normal sacrum (Peña, 2000).

2.3.3 Rectovaginal fistula

This is a rare malformation that affects only 1% of all cases. The rectum opens in to the vagina and the malformation could be low or high (Kiely, 1998).

2.3.4 Unusual female defects

Imperforate anus without a fistula is very rare in girls.

2.3.5 Persistent cloaca

The definition of this defect is that the rectum, vagina and urinary tract are fused together into a common channel. The length of this channel can vary but if the channel is longer than 3.5 cm the defect can be classified as a complex defect. It is not unusual to find varying degrees of septated or duplicated vagina and uterus (Peña, 2000). Associated urological defects appear in 90 % of girls with this high defect (Peña, 1990). Sacral defects and underdeveloped muscle complex is often seen.

2.3.6 Surgical treatment

A colostomy is generally done in a child with high or intermediate imperforate anus when the child is newborn. The colostomy provides decompression of the bowel and acts as protection for the final repair of the malformation (Peña, 2000). Posterior sagittal anorectoplasty (PSARP) means that the surgical incision is made between the patient's buttocks. The patient is placed in a prone position with the pelvis elevated. The external sphincter is located with electrical stimulation. The incision is then made exactly in the midline through all of the posterior musculature. The rectal pouch is

identified and the fistula is dissected and closed. The rectal pouch is mobilized and the different muscle layers are closed. The rectum is then placed with precision in the external sphincter and the skin is closed (Kiely, 1998; Peña, 1985, 2000).

The colostomy is kept for an additionally two to three months while the reconstructed anus is dilated with Hegar Dilator until the desired size is reached. The colostomy is then closed and the stools can pass through the neoanus.

2.3.6.1 MACE

MACE is a surgical method used as treatment for fecal incontinence. The appendix or an ileumconduit provide a continent catheterizable stoma and channel to the colon caecum. The patient takes a antegrade enema through the stoma daily in order to keep clean (Malone, Ransley & Kiely, 1990).

2.3.7 Functional outcome

Constipation and fecal incontinence are common problems in patients with high and intermediate imperforate anus. A study on children with high and intermediate imperforate anus showed that 35% of the patients had an excellent and 35% had a good bowel functional outcome (Rintala & Lindahl, 1995). Peña (2000) stated that of 1,192 patients 75% had voluntary bowel movements and 37.5% were totally continent. Another study showed that 96% of children with high and intermediate imperforate anus had a good or fair anorectal function (Tsuji et al., 2002).

2.4 CARE OF A CHILD WITH A CHRONIC CONDITION

Caring for a child involves protecting the child from danger and giving shelter and security. Children with anorectal malformations are a small but exposed group of children on which few studies have been made to study care both from a perspective of parent care and nursing care. Caring for a child with a malformation or a disability involves a lifelong responsibility for the child's well-being. Parents' participation in care and sometimes in treatment both at home and at the hospital are substantial (Hall, 2005a; Lam, Chang & Morrissey, 2005; Lawoko & Soares, 2004).

When the child is in need of hospital care the parents naturally want to stay at their child's bedside because of their commitment to care. Their feelings of love, protection, and responsibility and their desire to be involved in care make it obvious that the

parents should stay with their child (Dudley & Carr, 2004). Healthcare professionals often assume that parents really want to be engaged in the care of the child with malformations. Parents are therefore often obliged to act as nurses, performing complex clinical procedures on their children instead of being just a mother or a father (Kirk, Glendinning & Callery, 2005). Parents' daily care of their own children make them specialists; they observe small changes in the child's health condition that we as healthcare professionals may fail to see. Multiple dimensions of who really is the expert in care of a child with a chronic condition, the parents or the nurse, have to be considered when the child is in need of hospital care (Ford & Turner, 2001).

Parents' care of a child includes several aspects of physical, social, emotional, intellectual, spiritual, and developmental health. Healthcare professionals' care of the child involves clinical care to ensure physical health. One thing parents and healthcare professionals have in common, though is to return a sick child to health. This ambition can only be achieved by giving care from a family perspective as has been described by (Miceli & Clark, 2005).

Parental satisfaction will be achieved if the parents get involved in their child's care, and this can also decrease their feelings of stress. For this reason an open communication between healthcare professionals and parents is needed (Simons, 2002; Youngblut, Brooten & Kuluz, 2005). Sometimes communication between parents and healthcare professionals is somewhat limited, and a consequence of this might be that parents feel uninformed and insecure. Some studies have shown that nurses often underestimate parents' requests for information (Hall, 2005b; Sobo, 2004). In contrast, it has been found in a recently published study that parents of children with chronic conditions felt that they got more information than they could cope with (Hummelinck & Pollock, 2005).

2.5 PSYCHOSOCIAL ISSUES

Psychosocial function has different meanings and includes many different aspects. The social functioning of a child involves making friends, getting along with other children assimilating in groups, and participating in activities. However, the psychosocial concept could also include the child's ability to adjust socially and emotionally (Adams, Streisand, Zawacki & Joseph, 2002).

Imperforate anus and its consequences of constipation and fecal incontinence restrict a child in participating in social activities and may affect emotional adjustment. Parents' care of children during preschool years is extensive, but as the child grows up and starts school problems like incontinence will be more obvious and different psychosocial problems may come up. It has been found that among children with imperforate anus psychosocial problems, have been underestimated (Diseth, Emblem, Solbraa & Vandvik, 1994). Psychosocial problems such as secretive behavior among the children with imperforate anus to avoid teasing and bullying of classmates were described by (Ludman & Spitz, 1996).

It is difficult to compare imperforate anus with inflammatory bowel disease, but they do have very similar problems, such as the need for frequent toilet visits. A recently published paper reporting on psychosocial function and quality of life for children and adolescents with inflammatory bowel disease showed that these children and adolescents experienced a lower quality of life (De Boer, Grootenhuis, Derkx & Last, 2005). Another study showed normal psychosocial function among these children and adolescents but also showed that they had a larger amount of emotional/behavioral problems (Mackner & Crandall, 2005). Having a chronic condition, especially one of this kind, may result in psychosocial problems and affect the child's social contacts, since it is embarrassing to smell unpleasant.

Another study showed that depressive symptoms among children with chronic condition were related to the children's social relationships (Sandstrom & Schanberg, 2004). Pless and Nolan, (1991) also found that having a chronic illness increases the risk for psychosocial dysfunction. If a child has psychosocial problems it would probably affect the whole family. It has been found in an epidemiological study that childhood disability affects the entire family. In order to promote health and psychosocial functioning and to reduce stress symptoms, the family has to be seen as a whole (Witt, Riley & Coiro, 2003). Another study showed that parents of children with a chronic conditions experienced higher stress levels and felt more burdened by their families. This influenced the parent's well-being (Warschburger, Buchholz & Petermann, 2004).

Caring for a child with imperforate anus involves a lot of effort for the parents, and both the child and the parents may be affected by the consequences of the

malformation. The management of fecal incontinence is difficult, and its social consequences can be devastating. Our knowledge about these facts was limited and this is why this study was conducted.

3 AIMS

The overall aim of this thesis was to investigate experiences of children with high and intermediate imperforate anus and of their parents; how they have experienced care and treatment and what consequences the malformation involves for them.

Specific aim of included papers:

- I. The aim was to illuminate the experiences and psychosocial situation of 3 patients with high imperforate anus and their parents and to create a baseline for further research. (*Paper I*)
- II. The aim of the study was to evaluate the psychosocial effects of high and intermediate imperforate anus on school-aged children. (*Paper II*)
- III. The aim of this study was to evaluate parental experiences of care of children with high and intermediate imperforate anus. (*Paper III*)

4 DESIGN

This study is a product of a fully integrated cooperation between the pediatric surgery unit and the child and adolescent psychiatric unit. Such a close cooperation is needed if all aspects of nursing care for patients with chronic conditions and their families are to be incorporated.

This collaboration was developed as a common base for nursing care for patients at the St Görän Children's Hospital. We decided to focus on patients with imperforate anus because this malformation represents features that could be attached to both pediatric surgery and child and adolescent psychiatry. Private parts of the child's body are involved, and the consequences of the malformation and the follow up treatment include sensitive characteristics.

In a pediatric surgery perspective there had been considerations on how children with imperforate anus reacted on the malformation and the follow up treatment, but no further investigation had been performed at our clinic.

We choose to interview three selected patients with imperforate anus, who had severe problems with the malformation, and their parents. Our intention was to explore the experiences of living with imperforate anus and other factors to which it is related. This is similar to what is called an exploratory study, one that investigates the nature of the phenomenon, rather than simply observe and describes it (Polit & Beck, 2004).

The extensive finding from these interviews (*Paper I*) was the basis for questionnaires for children with high and intermediate imperforate anus and their parents. These questionnaires were used as study instruments in *Papers II* and *III*.

5 MATERIAL

5.1 MATERIAL PAPER I

Three patients with high imperforate anus and their families were interviewed. The patients were 12, 14 and 22 years old at the time of the interviews. There were three mothers and two fathers participating in the study. One father had left the family early on and was therefore not asked to participate since he had not been involved in the child's growing up. All three patients, suffered from a severe form of the malformation. Both the primary operation with a reconstruction of the anus and the primary follow-up treatment was done at St Göran Children's Hospital (Stockholm).

Inclusion criteria for participating in this study were; child born before 1987, high imperforate anus, problems with constipation and fecal incontinence, treatment with Malone's antegrade continence enema (MACE) and a willingness to discuss excretory problems they had experienced in childhood. Five children fulfilled the inclusion criteria and were contacted. Two of the patients did not want to take part in the study but three of them were willing to participate.

A stoma nurse first telephoned the families, briefly described the study, and asked them if they were willing to participate. After the children and parents gave their oral consent, they were sent written information about the study. Before the interviews started the family members got an information reminder about the purpose of the study to make them comfortable in talking about their experiences.

5.2 MATERIAL PAPERS II AND III

5.2.1 Children

Data in *Papers II* and *III* was obtained from 25 children with high or intermediate imperforate anus and their parents. The children had their definitive surgery for the anomaly with a PSARP performed at St. Göran Children's Hospital between 1987 and 1992. The original group consisted of 30 children; one of these patients had moved abroad and could not be located. The remaining 29 patients (9 boys and 20 girls) together with their parents were asked to participate in this study. They agreed but four families did not complete the study. The index group consisted of 25 children with high

or intermediate imperforate anus (9 boys and 16 girls). The mean age in the index group at the time of the study was 10.5 years (range 8.0 to 13.9).

Table 1. Types of defects in the Index Group

	Females	Males
	n=16	n=9
Persistent cloaca	5	
Rectovaginal fistula	1	
Rectovestibular fistula	10	
Rectovesical fistula		2
Rectoprostatic fistula		4
Rectobulbar fistula		3

Additional information about the child's extent of the lesion (Table 1) and bowel function at the time of the study was extracted from medical records and surgical reports. The majority of the children (19) had one or more additional congenital defects.

All of the children were operated with PSARP and fistula saving technique (Husberg, Lindahl, Rintala & Frenckner, 1992) by the same surgeon (B.F.) and the children had close and regular contact with the bowel therapist (M.Ö-J.). The children had regular follow-up treatment that included daily dilations with Hegar Dilators until the anorectum reached adequate size, and closure of the colostomy at the age of 6-12 months. Eleven children underwent dilations under anesthesia after the age of 18 months because of anal strictures. Malone's antegrade continence enema (MACE) was performed on five children because of problems with fecal incontinence.

Two groups of children similar to each other in age and gender distributions, along with their parents, served as controls. Control Group I consisted of 30 patients (5 boys and 25 girls) with Juvenile Chronic Arthritis (JCA) debuting before two years of age. The mean age was 10.6 years (range 7.8 to 13.6). Control Group II comprised 32 healthy (without chronic diseases) children (14 boys and 18 girls) who

were in the day surgery unit for a minor procedure, e.g. to repair a hernia. The mean age was 10.9 years (range 8.3 to 13.9).

5.2.2 Parents

Forty-five parents of children with high and intermediate imperforate anus participated in the index group. There were 25 mothers and 20 fathers in the group. Of the five missing fathers one was dead and the other four did not complete the study. Fifty-five parents of 30 children with juvenile chronic arthritis participated in Control Group I. There were 30 mothers and 25 fathers. Control Group II consisted of 52 parents of 32 healthy children. In this group there were 32 mothers and 20 fathers.

6 METHODS

6.1 INTERVIEWS, PAPER I

The interviews were conducted at the hospital by two of the authors in *Paper I*. Individual interviews were made by a nurse (M.N.) and a physician (P-A. R.). None of them had been involved earlier in the children's treatment or follow-up. The interview guide consisted of open-ended, and structured questions, and the sessions covered many different aspects of the child's malformation, treatment and the defect's psychosocial consequences. The parents and the patients described their own experiences of the situation. The interviews, between one and two hours long, were tape-recorded and transcribed.

6.1.1 Analyses of the interviews

The transcribed interviews were analyzed with a qualitative method, content analysis (Crabtree & Miller, 1999; Polit & Hungler, 1999). Two of the authors (M.N. and M.Ö-J.) read and reread the transcribed interviews. This was done to get a comprehensive impression of the text's contents. The two authors proceeded separately on the analyses with coding units, marked with different colours, according to content. These coding units, consisting of sentences or paragraphs, were grouped into categories identifying phenomena that emerged from the interviews (Morse & Field, 1998). This analysis was aimed at uncovering the meaning of the interview responses and finally these responses could be broken down into four categories based on the effects of the malformation: physical, emotional, social, and family.

6.2 QUESTIONNAIRES, PAPERS II AND III

The instruments used to gather data were questionnaires developed on the basis of the previous interview study. The results of the interviews showed which questions were of concern to the children and their parents and therefore essential to creating the self-structured questionnaires. The children's and parents' questions were mostly congruent. The questionnaires were tested on both children and their parents in three subsequent pilot studies. Minor revisions of the questionnaires were made after each pilot study and eventually they were completed.

6.2.1 Children's questionnaire

The children's questionnaire consisted of 45 items that were divided into five psychosocial domains (school, sport activities, peer relationships, self-esteem, and emotional health) and one physical domain. The items were scored on a five-point Likert scale. The first part of the questionnaire was designed as a pictogram that featured five faces with various expressions. The alternative answers were: very sad, rather sad, neither sad nor happy, rather happy, and very happy. This first section contained 24 questions. The final section lasted of 21 items with five alternative answers: always, almost always, sometimes, almost never, and never. These items were labeled with words to clarify the meaning of the scale points.

6.2.2 Parents' questionnaire

The parent's questionnaire consisted of 59 items covering several domains such as the child's hospital care, psychosocial issues concerning both the child and the parent, and finally the child's physical functioning associated with the malformation. The items were scored on a visual analogue scale (VAS). All the items had a VAS ranking from 0-100. Each question had two anchor words, i.e. negative - positive, never - always, very bad - very good.

6.2.3 Statistical analyses of the questionnaires

The data were analyzed using the STATA (version 7) program for statistical analysis.

6.2.3.1 Children's questionnaire

The children's questionnaires were analyzed with Pearson's X^2 -square test and the Fisher's exact test for statistical intergroup differences. Ordered logistic regression was used to analyze the relationship between variables and the psychosocial outcome. The index group was compared with one control group at a time. A p -value of less than .05 was considered statistically significant.

6.2.3.2 Parents questionnaire

Parents' questionnaires were analyzed with a t-test and the Wilcoxon Rank-Sum Test for statistical intergroup differences. Differences between mothers' and fathers' answers on the pair level were analyzed with the Wilcoxon Signed Rank Test. The index group was compared to one control group at a time. A p -value of less than .05 was considered statistically significant.

6.3 CHILD BEHAVIOR CHECKLIST

Swedish versions of the Child Behavior Checklist (CBCL/4-18) (Achenbach, 1991a) and the Teacher's Report Form (TRF) (Achenbach, 1991b) were used to measure children's emotional and behavioral problems as reported by parents and teachers. The CBCL presents descriptions of 120 emotional and behavioral problems that are likely to occur in children 4 to 18 years of age. The CBCL yields a total behavior problem score and two sub-scales referred to as internalizing (social, withdrawal, depression, anxiety) and externalizing (antisocial, aggressive, delinquency) symptom scales. The CBCL includes items about social competence, which measure the child's social ability and capacity in school and during extracurricular activities and hobbies.

6.3.1 Statistical analyses of CBCL and TRF

The data were analyzed using the STATA (version 7) program for statistical analysis. Data from the CBCL and TRF were analyzed according to instructions in the respective manuals (Achenbach, 1991a, 1991b).

6.4 ETHICAL CONSIDERATIONS

The local ethical committee at Karolinska Hospital approved these studies (Reference Number 99-202). The study was introduced to children and their parents both through a verbal presentation and by providing them with written information. Informed consent was received from all respondents.

7 RESULTS

7.1 PAPER I

Four categories of effects of the malformation emerged from the analyses of the interviews: physical, emotional, social, and family. The physical effects of the child's malformation influenced the family's experiences and permeated the interviews. These effects were linked with the other categories and were therefore not treated as an entity. The emotional effects of the malformation were the most frequently occurring effect in the data. The social effects described how the family's daily life was affected socially, and the family effects dealt with the how family members related to one another.

7.1.1 Physical effects of the malformation

The physical effects of imperforate anus were apparent from the very first hour of the child's life, in the delivery ward. A colostomy had to be created during the first days, as the baby could not pass stools normally. A reconstruction of the anus was subsequently performed, and the parents had to dilate the anus to optimal opening width for several months. To prevent constipation and fecal incontinence, the children were given enemas every other day for several years. The children had to wear diapers or other protection against soiling themselves until reached preschool age. Incontinence also involved fecal odor. After several years of these difficulties, the children were offered a MACE operation.

7.1.2 Emotional effects of the malformation

Sorrow, pain, and disappointment were the most frequently described emotional effects of the malformation. These emotions were closely connected with anal manipulations (dilations and enemas) and incontinence. Parents felt that they had to force themselves on the child when they carried out the dilations, and that it became a fight. As the child grew older one parent had to hold him/her down, while the other parent performed the dilatation. All the parents stressed that they could never get used to it. The children themselves did not remember the dilations and how it had felt. The parents stated that prevention of constipation and incontinence was even worse than dilations. The children gave only very brief answers on this question. Disappointment and anger among the parents appeared when they talked about the reconstruction of the anus and

the child's bowel function. Toilet habits became very central in their lives for a long time. The parents were disappointed with information about the consequences of the malformation.

The emotional effect of sorrow was found when the parents reported that their children had experienced periods of depression. All three children acknowledged that they had been depressed. Feelings of shame are concealed, but recognizable when the parents talked about the fecal incontinence. Feelings of fear are apparent when parents discuss the children's fright and pain in reaction to anal dilations. The positive emotional effects of satisfaction and joy are visible when parents and children discuss the MACE operation, which effected a positive change.

7.1.3 Social effects of the malformation

Procedures to prevent constipation and soiling interfered with the family's activities. Enemas and washouts were demanding and time-consuming and had to be planned with accuracy. Sports participation and the showers afterwards had been difficult. Bullying had occurred.

7.1.4 Family effects of the malformation

Relationships within families were strained from time to time because of the malformation. One mother told of quarrels over how to handle the treatment of the child. There were also disagreements about how to handle the enemas. The mothers said that they bore the main responsibility for everyday care; they did most of the dilations and enemas. The fathers and the children confirmed this.

7.2 PAPER II

Children with imperforate anus indicated an overall positive attitude and reported that they enjoyed school, had good relationship with schoolmates, had several friends, and participated in leisure activities. The children with imperforate anus said they were able to express their will and stated that they were generally in a good mood. Constipation occurred more frequently than in control groups. There were no correlations between the severity of the original malformation and psychosocial outcome, nor was any correlation found between MACE and psychosocial outcome. Overall, no gender differences were found.

Parents of children with imperforate anus recorded more emotional and behavioral problems on the CBCL questionnaire than did parents of children in the control groups. Teachers recorded few emotional and behavioral problems among children with imperforate anus.

7.2.1 School

Comparing variables related to school activities between the children with imperforate anus and the children in the two control groups, a small number of statistically significant differences were found in favor of the index group. The children liked school more ($p<.05$) compared to children in Control Group I, and they reported having better relationships with their classmates ($p<.05$) compared with Control Group II. The parents confirmed these findings.

Children in the index group did not like using the school toilet for defecation and a statistically significant difference was found compared with Control Group I ($p<.05$). None of the children with imperforate anus reported being bullied but they had been teased. School breaks were viewed positively by the children. Physical education in school and showering afterwards was also considered positive.

7.2.2 Sports activities and hobbies

No statistically significant difference was found between groups regarding membership on a sports team. In the index group 84% of the children participated in some form of sport activity. In Control Group I 85% of the children participated in sports and in Control Group II 90% of children participated in sport activities. Children in all groups had one or more hobbies during leisure time.

7.2.3 Peer relationships

Children with imperforate anus reported that they had friends and that they had good relationship with their friends. They reported that they were more appreciated by their friends than did children in Control Group II ($p<.01$). They could also make decisions in a group just as often as children in the two control groups. No statistically significant difference was found between the groups in having a best friend and spending a lot of time with their best friend. Twenty (83%) of the children with imperforate anus, 28

(93%) of the children with JCA, and 22 (71%) of the healthy children reported that they had a best friend.

7.2.4 Emotional health

Children's emotional health and self-esteem scores did not differ between groups. The children with imperforate anus reported that they liked themselves and their bodies, and reported less frequent thoughts ($p < .05$) about their bodies compared with their healthy peers. Children in all groups thought of their problems now and then but they estimated their parent's worries about their inconvenience as high. Both the index group and the two Control Groups stated that they were very much loved by their parents.

7.2.5 Physical function

Constipation was most frequently reported in the group of children with imperforate anus. Eighteen (72%) of the children with imperforate anus said that they were "always" or "almost always" constipated compared with four (13%) of the children with JCA, and four (13%) of the children in the healthy group. In the index group the mothers' assessments of the child's bowel function showed statistically significant differences on constipation, fecal leakage and fecal odors compared with the control groups.

7.2.6 CBCL: parents' and teachers' report

On the CBCL total problem score scale a statistically significant difference was found between the index group and Control Group I ($p < .05$). However, sub-scales for internalizing and externalizing evinced no significant differences between groups. Teachers' reports on the TRF did not differ between groups.

7.3 PAPER III

7.3.1 Care

Statistical significant differences were found between parents in the index group and parents in the Control Groups I ($p < .01$) and II ($p < .05$) in mothers' reports on how they had been taken care of in the hospital. A statistically significant difference was found between the fathers in the index group and Control Group I ($p < .05$) on how they had been taken care of in the hospital. Both mothers and fathers in the index group were less satisfied with how their children had been taken care of during their hospital stay,

and a statistically significant difference was found between mothers and fathers of children with imperforate anus compared to mothers and fathers in Control Group I ($p < .01$).

The mothers in the index group reported less understanding and more negative experiences of the treatment, and statistically significant differences were found compared to both Control Groups. Fathers of children with imperforate anus also reported more negative experiences of their child's treatment and a statistically significant difference was found compared to fathers in Control Group II. The children's comprehensions of treatment, as reported by their parents, were lower in the index group compared with the two control groups.

7.3.2 Information

Satisfaction with information about the child's treatment was lower among fathers in the index group, and a statistically significant difference was found compared to fathers in Control Group II. Information to the child about treatment as reported by the mothers and the fathers in the index group was not satisfactory. A statistically significant difference was found compared to mothers in Control Group I and to fathers in Control Group II. Advice about the child's difficulties from the hospital was not sufficient as reported by the parents in the index group, and statistically significant differences were found compared to mothers and fathers in Control Groups I and II.

7.3.3 Involvement

Mothers and fathers in the index group reported their involvement in the child's care to be high, and statistically significant differences were found between fathers in the index group compared to the fathers in Control Group I. Children's involvement in the care was low as reported by their parents. Statistically significant differences between groups were found compared with mothers in Control Groups I and II and fathers in Control Group II.

7.3.4 Differences on the pair level

In the index group there were no statistically significant differences between the mothers' and fathers' answers on questions about care, information, and involvement on the pair level.

8 DISCUSSION

8.1 METHODOLOGICAL CONSIDERATIONS

8.1.1 Paper I

In an interview study the respondents are given the opportunity to tell their story, and sometimes it can become more of a therapeutic conversation. The respondents have the opportunity to tell as much as they want about an issue, and the parents in *Paper I* gave long narrations about their experiences. A large amount of text has to be compressed in to smaller parts, and this was done with a qualitative analysis (Kvale, 1996). In both qualitative and quantitative analysis the aim is to organize, provide structure for, and elicit meaning from research data. In the qualitative content analysis the researcher acts as an interpreter, reading through the data looking for meaningful segments and units and developing a categorization scheme (Polit-O'Hara, Hungler & Polit, 1997). It is a challenge in the qualitative analysis for the researcher to make some sense of all the transcribed unstructured data as it consists of many pages of text. Silverman, (2001) describes methods of strengthening the trustworthiness of a qualitative study. This can be improved by comparing the analysis of the same data by several authors, and this was done with data in *Paper I*. The respondents in *Paper I* received copies of the interview guide after the interviews and were given the possibility to give us comments on it. None of the respondents sent the interview guide back, and when we called them on the telephone none of them had anything to comment on or add from the interview guide. The results from a qualitative study are always subjective and cannot be generalized to apply to other groups, although they bring a deeper understanding of a specific phenomenon.

8.1.2 Papers II and III

In *Papers II* and *III* questionnaires were used. All data were collected at one and the same time and the study had a design for comparing groups. In quantitative research when the phenomena of health or psychosocial functioning are studied, questionnaires are often used as an instrument.

This sample of children is rather small, but still it was the total amount of children with high and intermediate imperforate anus born 1987-1992 and treated in Stockholm. This

small sample may of course affect the results and make it difficult to draw firm conclusions.

The children's questionnaire was designed to be easy for the children to answer and the Likert scale we used has been found to be easy for children to complete (van Laerhoven, van der Zaag-Loonen & Derkx, 2004). Two studies found that children younger than 6 years often give more extreme answers than do 7-9 years-old children on questions of an emotional type (Chambers & Johnston, 2002; Rebok et al., 2001). The children in our study were 8-13.9 years old and this might give strength to the children's answers. Rebok et al., (2001) also reported that children at the age of 8 were able to report on virtually presented questions about their health. One additional thing that could give justification for developing a specific children's questionnaire is that all disease-specific quality of life instruments for patients with fecal incontinence are developed for adults and are inappropriate for use with children. These questionnaires contain issues that are irrelevant to ask children about (Trajanovska & Catto-Smith, 2005).

The parents' questions were developed to correspond to the children's questions, in order to look for differences and similarities between parents and children. In the parents' questionnaire a visual analogue scale (VAS) was used. The questionnaire had many questions and answering on a VAS has been found to be easy to handle. VAS is often used to assess pain, but it has also been used in studies of health outcome (Lukacz et al., 2004). The VAS in the parents' questionnaire in our study had two anchor words; this is preferable and makes it easier for the respondents to fill in the questionnaire (Torrance, Feeny & Furlong, 2001). The CBCL is a standardized instrument and is frequently used in research of behavior/emotional problems (Achenbach, 1991a, 1991b). It is a proxy version and it reflects the parent's assessment of the child's psychosocial function.

8.2 REFLECTIONS ON THE FINDINGS

In *Paper I* four categories - physical, emotional, social and family effects - emerged from the interviews. The parents' distress was overwhelming, with painful memories of caring for their children and all the things they had been forced to deal with. In general, emotions were the most recurrently mentioned effect of the malformation, and negative emotions like suffering, sorrow, and pain were the emotions most frequently reported

emotions by the parents. Positive emotions like joy and satisfaction were mentioned in relation to the MACE operation.

The suffering that parents in *Paper I* experienced was a great surprise to us, although we knew that some children with imperforate anus have many problems with constipation and incontinence. However, we as healthcare professionals could not imagine that caring for these problems at home could be so extensive, and by doing these interviews we discovered that our knowledge about caring for a child with imperforate anus was limited. It seemed as if these families' lives had revolved entirely around this concern. It has been found in other studies on parents of children with other chronic conditions and malformations that there is an increased risk for parenting stress and mixed feelings of happiness, despair, and guilt in such situations (Erickson & Ray, 2004; Johansson & Ringsberg, 2004; Streisand, Swift, Wickmark, Chen, & Holmes, 2005; Uzark & Jones, 2003).

The parental suffering and disappointment described in *Paper I* are also discernable in *Paper III*. In *Paper II* the children with imperforate anus seemed to be happy and unworried about their inconvenience when they reported by themselves, though their parents have higher scores for more emotional/behavioral problems in CBCL. This might be a reflection of the parents' view about the child's inconvenience.

The parental satisfaction with care, involvement, and information in *Paper III* was lower among parents of children with imperforate anus than in the control groups. Having a child with a malformation is stressful (Ludman & Spitz, 1996) for the parents and their own feelings of guilt because of the child's defect might be an explanation for our finding. Hassink et al., (1998) found that having a child with an anorectal malformation does not always involve problems, but certain groups of parents could be more at risk with respect to parenting stress. The parents are in need of extra support in caring for the child.

The need of psychological support might have been underestimated among children with imperforate anus and their parents. This is obvious in *Paper I* where the parents did not seem to have talked about their experiences. When the child has a chronic condition, parents often experience emotional disturbance like anger and guilt. It is thus essential that the family be seen as a whole (Hopia et al., 2005).

Support and advice about the malformation may be given to some extent by the bowel therapist and the pediatric surgeon but if the problems change character to more psychopathology, then psychiatric help is required. Imperforate anus is a malformation that concerns a very sensitive part of the body and to help these families on a psychological level, extensive knowledge about the malformation is essential. To avoid misunderstandings and to avoid the risk of accusing the parents of something horrible all professionals involved in care of children with imperforate anus must know that the follow-up treatment could be experienced by the parents as an act of abuse.

The children/adolescents in *Paper I* had their anal reconstruction done before 1987 and were operated on with another technique than that used today. Since 1987 all children with imperforate anus have been are operated on with PSARP and the fistula-saving technique in order to preserve the anlage of the internal sphincter. This saving of the internal sphincter anlage may preserve a better sphincter function than with earlier reconstructions of the anus although it is not realistic to think that children with imperforate anus will attain a normal function of the internal sphincter (Husberg et al., 1992). The children in *Paper II* have all had the PSARP when they were very young, about six months old, and the follow-up treatment with dilations were done before the age of one year.

Follow-up studies done on children operated with the PSARP and the fistula-saving technique demonstrates that the patients seem to achieve an acceptable continence (Liu, Yuan, Geng, Wang & Li, 2004; Rintala & Lindahl, 2001; Tsuji et al., 2002). Since 1987 a stricter schedule with a homogenous surgical treatment and follow-up of children with imperforate anus has been followed at our hospital. The children and their parents now regularly meet the bowel therapist between visits to the pediatric surgeon. Parents and children in *Paper II* may have had an earlier and better support from the hospital than did the parents and children in *Paper I*.

It seems as if the children in *Paper II* have a better bowel function than patients in *Paper I* do, even though we do not have any objective assessment of that. Children in *Paper I* and *II* are not comparable since they have been operated with diverse surgical methods, and there is also a difference in age. The children in *Paper II* had not yet reached adolescence, when additional issues as sexual identity become important. The adolescents in *Paper I* had difficulties in controlling their bowel movements, and they

have had severe problems with constipation and fecal incontinence. The odor that fecal incontinence entails is easily revealed by others. Not being able to have the control over one's own body could result in a socially isolated behaviour among adolescents (van Kuyk et al., 2001).

The children in *Paper I* had been exposed to teasing and bullying. This malformation, with the inconveniences it brings, can stigmatize the child and make him/her an object of ridicule. However, in *Paper II* the children admitted being teased, but every child in the index group denied being bullied. Children's ways of coping with difficult situations can involve denial both to themselves and to others. Denial is a psychologically protective mechanism (Klennert, McQuaid, McCormick, Adinoff & Bryant, 2000; Ludman & Spitz, 1996).

Since this malformation appears when the children are born, they are treated at a clinic for pediatric surgery in the beginning. The children are also followed up during their childhood and adolescence in the pediatric surgery clinic. However, in the transition into adulthood the changeover to care in an adult clinic can be a big step (Little et al., 2003). Colorectal surgery units do not always have the same expertise and knowledge of what consequences this specific malformation entails. Hassink, Rieu, Severijnen, Brugman-Boezeman and Festen, (1996) found in their study that it was a problem for adult patients with imperforate anus to get accurate advice on how to handle constipation and fecal incontinence. Since this treatment has improved in recent years the adult patients might not be sufficiently informed, and this was seen by Hartman et al., (2004) in a study on adult patients with imperforate anus in Holland. MACE has been one of the most successful treatments of fecal incontinence for patients born with imperforate anus and in *Paper I* parents and children are very positive when they mention MACE. With this surgical method patients with fecal incontinence can achieve social continence (Aksnes et al., 2002; Curry, Osborne & Malone, 1998; Malone et al., 1990; Meier, Foster, Guzzetta & Coln, 1998).

Parents experienced that they did not get enough information. Descriptions of the advanced methods of pediatric surgery that are used to repair this complicated malformation are not always so easy to understand, even though information is presented in a proper way. It has been found that a simply written information sheet and a drawing are often the best way for the surgeon to give information about an advanced

surgical operation (Noll, Spitz & Pierro, 2001). A specialist nurse could also be of assistance in presenting this important information.

The parents often seek more information than is given to them at the hospital, and an alternative resource for information about this unusual malformation is the use of the Internet. The medical guidance parents will get by searching on the Internet is not always optimal, as many websites are of poor quality. Even so, Internet information will provide parents with new facts, though it may not always give a better understanding (Aslam, Bowyer, Wainwright, Theologis & Benson, 2005; Boston, Ruwe, Duggins & Willging, 2005; Impicciatore, Pandolfini, Casella & Bonati, 1997).

Parents seem to have been very much involved in their children's treatment and have had tough experiences of the child's malformation (*Paper I* and *Paper III*). Parents of children with a chronic condition may gain support by meeting other parents in the same situation. It has been found that that family-to-family support can have important psychological benefits, above all for the mothers since they are often the main caregivers (Ireys, Chernoff, DeVet & Kim, 2001). Those parents of children with a chronic condition who participate in parents' groups increase their own knowledge, get a better understanding about other families' concerns, and may experience empowerment (Young, McMenemy & Perrin, 2001). Barlow, Coren and Stewart-Brown, (2002) described that even among healthy children a parenting program could improve the maternal psychosocial health. The use of Internet, apart from providing parents with information, can also serve as a parental support since parents meet other parents on different websites (Sarkadi & Bremberg, 2005).

The children did not report any psychosocial impairment (*Paper II*). The children had some leakage and fecal odor, as reported by their parents. The children were positive about school and had friends and the parents confirmed their children's satisfaction about school and social activities. These positive findings were in contrast to those on children in *Paper I*, who admitted that they had sometimes felt depressed. Support networks for adolescents with a chronic condition are important for sharing experiences. Kyngäs, (2004) found that in such a network many different supporters exist, including parent's, peers, school, healthcare providers, technology, and in some cases even a pet. An additional way to help the adolescents cope with their chronic

condition could be to let them participate in their own care by involving them in medical decision-making (Schmidt, Petersen & Bullinger, 2003).

The parents have recorded more emotional and behavioral problems on CBCL compared with the control groups (*Paper II*). However the teachers seemed to think that children with imperforate anus did not differ from their classmates. This finding is in line with other studies on psychosocial functioning and quality of life among patients with imperforate anus and other chronic conditions, where no difference between them and healthy peers could be observed (Diseth, Bjornland, Novik & Emblem, 1997; Ditesheim & Templeton, 1987; Hartman et al., 2004; Hassink, Rieu, Brugman & Festen, 1994; Mackner & Crandall, 2005).

Parents with all their knowledge about the malformation and the consequences may observe their children in a different way. In school, though, other qualities among the child are noticed like learning, ability to concentrate, and social competence among classmates in breaks. The school environment is important to children and these children seem to have a lot of support from teachers and other adults to help the children in handling their problems.

Children in *Paper II* were between 8-13 years old (mean age 10.5 years) when they participated in the study. At this age the parents are still the most important people in a child's life, handling the practical things like preparing their children for different events. Social relationships with friends and classmates are important for the child. As this study has shown, it seems as though the parents support their children in a manner that helps them to avoid accidents with fecal incontinence and odor in the school situation. This malformation does not show on the outside as a malformed arm or leg would, but consequences of incontinence can become very obvious because of the odor. In a recently published study it was found that fecal incontinence is a social handicap and that there was a higher risk of depression among children with imperforate anus as the children grew older and moved into adolescence (Funakosi et al., 2005). Another study found that fecal incontinence and frequent visits to the toilet could be one of the main factors compromising quality of life (Ceriati et al., 2004).

The findings in *Paper III*, showing no differences in answers on care from mothers and fathers, may be due to the fact that parents filled in the questionnaire together, or that

they share the same opinions about the children's care. These parents have dealt with their children's difficulties for several years, and the parents have probably developed some kind of teamwork for handling the children's inconveniences in order to maintain as normal a family life as possible.

There was no difference in mothers' and fathers' answers on distribution of responsibility for care of the children. This can be interpreted to mean that they have both taken a great responsibility (*Paper III*). This finding was in contrast to what the mothers in the interviews reported in *Paper I*. A mother often takes a great responsibility for a child with a chronic condition, but if the father is involved in the daily care he establishes a better relationship with their child. This can ease the mother's burden (Roach, Orsmond & Barratt, 1999). A benefit for the entire family could be to strengthen the father. In a recently published study on fathers parenting children with a chronic condition, it was found that they expressed concern for both their children and their spouses (Hovey, 2005). Caring for a child with imperforate anus puts an extra strain on the family. It has been found that having a child with a chronic condition can affect family members' own well-being, their relationships with each other, and with outside world (Hopia et al., 2005).

Papers I, II and III show that our knowledge about children with imperforate anus is still not sufficient, even though we meet these children and their parents daily in our work at the pediatric surgery clinic. This malformation is unique, and the consequences it entails can be embarrassing and shameful. If we as specialists caring for children with imperforate anus invite the parents and the child to a discussion meeting, hopefully they will share their experiences with us. This will increase our knowledge and enable us to offer the best of resources. The bowel therapist could give psychosocial support, and when needed can be a liaison to the child and adolescent psychiatry unit. The amount of psychosocial support for these children and their parents has been limited earlier in our clinic. A liaison between the pediatric surgery unit and the child and the adolescent psychiatry unit is needed, and should therefore be a natural part of our work.

9 CONCLUSIONS

- Parents of children with high imperforate anus had experienced suffering and many difficulties associated with the malformation.
- Children with imperforate anus in this study did not seem to have any psychosocial impairment, though their parents stated that they experienced leakage, constipation, and fecal odor.
- The children might have some psychosocial difficulties according to parents, but not according to their teachers.
- Parents of children with imperforate anus experienced less satisfaction with medical care the children had received, and reported that support had been insufficient.

Care of a child with imperforate anus entails intricate situations both for the children and for their parents. An open communication and an interaction between parents and healthcare professionals are important. Psychosocial support for children with imperforate anus and their parents has been limited in our clinic earlier, and a liaison between the pediatric surgery unit and the child and adolescent psychiatry unit would be desirable.

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11 REFERENCES

- Achenbach,T.M. (1991a). *Manual for the child behavior checklist/4-18 and 1991 profile*. Burlington, VT: Dept. of Psychiatry, University of Vermont.
- Achenbach,T.M. (1991b). *Manual for the Teacher's report form and 1991 profile*. Burlington, Vt.: Dept. of Psychiatry, University of Vermont.
- Adams,C.D., Streisand,R.M., Zawacki,T., & Joseph,K.E. (2002). Living with a chronic illness: a measure of social functioning for children and adolescents. *J Pediatr Psychol*, 27(7), 593-605.
- Aksnes,G., Diseth,T.H., Helseth,A., Edwin,B., Stange,M., Aafos,G., et al. (2002). Appendicostomy for antegrade enema: effects on somatic and psychosocial functioning in children with myelomeningocele. *Pediatrics*, 109(3), 484-489.
- Aslam,N., Bowyer,D., Wainwright,A., Theologis,T., & Benson,M. (2005). Evaluation of Internet use by paediatric orthopaedic outpatients and the quality of information available. *J Pediatr Orthop B*, 14(2), 129-133.
- Barlow,J., Coren,E., & Stewart-Brown,S. (2002). Meta-analysis of the effectiveness of parenting programmes in improving maternal psychosocial health. *Br J Gen Pract*, 52(476), 223-233.
- Boston,M., Ruwe,E., Duggins,A., & Willging,J.P. (2005). Internet use by parents of children undergoing outpatient otolaryngology procedures. *Arch Otolaryngol Head Neck Surg*, 131(8), 719-722.
- Ceriatì,E., Deganello,F., De Peppo,F., Ciprandi,G., Silveri,M., Marchetti,P., et al. (2004). Surgery for ulcerative colitis in pediatric patients: functional results of 10-year follow-up with straight endorectal pull-through. *Pediatr Surg Int*, 20(8), 573-578.
- Chambers,C.T., & Johnston,C. (2002). Developmental differences in children's use of rating scales. *J Pediatr Psychol*, 27(1), 27-36.
- Crabtree,B.F., & Miller,W.L. (1999). *Doing qualitative research* (2. ed.). Thousand Oaks, Calif.: SAGE.
- Curry,J.I., Osborne,A., & Malone,P.S. (1998). How to achieve a successful Malone antegrade continence enema. *J Pediatr Surg*, 33(1), 138-141.
- De Boer,M., Grootenhuis,M., Derkx,B., & Last,B. (2005). Health-related quality of life and psychosocial functioning of adolescents with inflammatory bowel disease. *Inflamm Bowel Dis*, 11(4), 400-406.
- Diseth,T.H., Bjornland,K., Novik,T.S., & Emblem,R. (1997). Bowel function, mental health, and psychosocial function in adolescents with Hirschsprung's disease. *Arch Dis Child*, 76(2), 100-106.
- Diseth,T.H., Emblem,R., Solbraa,I.B., & Vandvik,I.H. (1994). A psychosocial follow-up of ten adolescents with low anorectal malformation. *Acta Paediatr*, 83(2), 216-221.
- Ditesheim,J.A., & Templeton,J.M., Jr. (1987). Short-term v long-term quality of life in children following repair of high imperforate anus. *J Pediatr Surg*, 22(7), 581-587.

- Dudley,S.K., & Carr,J.M. (2004). Vigilance: the experience of parents staying at the bedside of hospitalized children. *J Pediatr Nurs*, 19(4), 267-275.
- Erickson,D.V., & Ray,L.D. (2004). Children with chronic continence problems: the challenges for families. *J Wound Ostomy Continence Nurs*, 31(4), 215-222.
- Ford,K., & Turner,D. (2001). Stories seldom told: paediatric nurses' experiences of caring for hospitalized children with special needs and their families. *J Adv Nurs*, 33(3), 288-295.
- Funakosi,S., Hayashi,J., Kamiyama,T., Ueno,T., Ishii,T., Wada,M., et al. (2005). Psychosocial liaison-consultation for the children who have undergone repair of imperforate anus and Hirschsprung disease. *J Pediatr Surg*, 40(7), 1156-1162.
- Hall,E. O. (2005a). Being in an alien world: Danish parents' lived experiences when a newborn or small child is critically ill. *Scand J Caring Sci*, 19(3), 179-185.
- Hall,E. O. (2005b). Danish parents' experiences when their new born or critically ill small child is transferred to the PICU-a qualitative study. *Nurs Crit Care*, 10(2), 90-97.
- Hartman,E.E., Oort,F.J., Aronson,D.C., Hanneman,M.J., van der Zee,D.C., Rieu,P.N., et al. (2004). Critical factors affecting quality of life of adult patients with anorectal malformations or Hirschsprung's disease. *Am J Gastroenterol*, 99(5), 907-913.
- Hassink,E.A., Brugman-Boezeman,A.T., Robbroeckx,L.M., Rieu,P.N., van Kuyk,E.M., Wels,P.M., et al. (1998). Parenting children with anorectal malformations: implications and experiences. *Pediatr Surg Int*, 13(5-6), 377-383.
- Hassink,E.A., Rieu,P.N., Brugman,A.T., & Festen,C. (1994). Quality of life after operatively corrected high anorectal malformation: a long-term follow-up study of patients aged 18 years and older. *J Pediatr Surg*, 29(6), 773-776.
- Hassink,E.A., Rieu,P.N., Severijnen,R.S., Brugman-Boezeman,A.T., & Festen,C. (1996). Adults born with high anorectal atresia--how do they manage? *Dis Colon Rectum*, 39(6), 695-699.
- Hopia,H., Tomlinson,P.S., Paavilainen,E., & Astedt-Kurki,P. (2005). Child in hospital: family experiences and expectations of how nurses can promote family health. *J Clin Nurs*, 14(2), 212-222.
- Hovey,J.K. (2005). Fathers parenting chronically ill children: concerns and coping strategies. *Issues Compr Pediatr Nurs*, 28(2), 83-95.
- Hummelinck,A., & Pollock,K. (2005). Parents' information needs about the treatment of their chronically ill child: A qualitative study. *Patient Educ Couns*.
- Husberg,B., Lindahl,H., Rintala,R., & Frenckner,B. (1992). High and intermediate imperforate anus: results after surgical correction with special respect to internal sphincter function. *J Pediatr Surg*, 27(2), 185-188; discussion 188-189.
- Impicciatore,P., Pandolfini,C., Casella,N., & Bonati,M. (1997). Reliability of health information for the public on the World Wide Web: systematic survey of advice on managing fever in children at home. *Bmj*, 314(7098), 1875-1879.
- Ireys,H.T., Chernoff,R., DeVet,K.A., & Kim,Y. (2001). Maternal outcomes of a randomized controlled trial of a community-based support program for families of children with chronic illnesses. *Arch Pediatr Adolesc Med*, 155(7), 771-777.

- Johansson,B., & Ringsberg, K.C. (2004). Parents' experiences of having a child with cleft lip and palate. *J Adv Nurs*, 47(2), 165-173.
- Kiely,E.M., Peña, Alberto. (1998). Anorectal Malformations. In J. A. O'Neill (Ed.), *Pediatric surgery* (5th ed., Vol. 2 vol.). St. Louis ; London: Mosby.
- Kirk,S., Glendinning,C., & Callery,P. (2005). Parent or nurse? The experience of being the parent of a technology-dependent child. *J Adv Nurs*, 51(5), 456-464.
- Klennert,M.D., McQuaid,E.L., McCormick,D., Adinoff,A.D., & Bryant,N.E. (2000). A multimethod assessment of behavioral and emotional adjustment in children with asthma. *J Pediatr Psychol*, 25(1), 35-46.
- Kvale,S. (1996). *Interviews: an introduction to qualitative research interviewing*. Thousand Oaks, Calif.: Sage.
- Kyngäs,H. (2004). Support network of adolescents with chronic disease: adolescents' perspective. *Nurs Health Sci*, 6(4), 287-293.
- Lam,L.W., Chang,A.M., & Morrissey,J. (2005). Parents' experiences of participation in the care of hospitalised children: A qualitative study. *Int J Nurs Stud*.
- Lawoko,S., & Soares,J.J. (2004). Satisfaction with care: a study of parents of children with congenital heart disease and parents of children with other diseases. *Scand J Caring Sci*, 18(1), 90-102.
- Little,D.C., Rescorla,F.J., Grosfeld,J.L., West,K.W., Scherer,L.R., & Engum,S.A. (2003). Long-term analysis of children with esophageal atresia and tracheoesophageal fistula. *J Pediatr Surg*, 38(6), 852-856.
- Liu,G., Yuan,J., Geng,J., Wang,C., & Li,T. (2004). The treatment of high and intermediate anorectal malformations: one stage or three procedures? *J Pediatr Surg*, 39(10), 1466-1471.
- Ludman, L., & Spitz,L. (1996). Coping strategies of children with faecal incontinence. *J Pediatr Surg*, 31(4), 563-567.
- Lukacz,E.S., Lawrence,J.M., Burchette,R.J., Luber,K.M., Nager,C.W., & Buckwalter,J.G. (2004). The use of Visual Analog Scale in urogynecologic research: a psychometric evaluation. *Am J Obstet Gynecol*, 191(1), 165-170.
- Mackner,L.M., & Crandall,W.V. (2005). Long-term psychosocial outcomes reported by children and adolescents with inflammatory bowel disease. *Am J Gastroenterol*, 100(6), 1386-1392.
- Malone,P.S., Ransley,P.G., & Kiely,E.M. (1990). Preliminary report: the antegrade continence enema. *Lancet*, 336(8725), 1217-1218.
- Meier,D.E., Foster,M.E., Guzzetta,P.C., & Coln,D. (1998). Antegrade continent enema management of chronic fecal incontinence in children. *J Pediatr Surg*, 33(7), 1149-1151; discussion 1151-1142.
- Miceli,P.J., & Clark,P.A. (2005). Your patient--my child: seven priorities for improving pediatric care from the parent's perspective. *J Nurs Care Qual*, 20(1), 43-53; quiz 54-45.
- Morse,J.M., & Field,P.A. (1998). *Nursing research: the application of qualitative approaches* (2. ed.). Cheltenham: Stanley Thornes.

- Noll,S., Spitz,L., & Pierro,A. (2001). Additional medical information: prevalence, source, and benefit to parents. *J Pediatr Surg*, 36(5), 791-794.
- Peña,A. (1985). Surgical treatment of high imperforate anus. *World J Surg*, 9(2), 236-243.
- Peña,A. (1990). *Atlas of surgical management of anorectal malformations*. New York: Springer-Verlag.
- Peña,A. (2000). Imperforate Anus And Cloacal Malformations. In K. W. Ashcraft (Ed.), *Pediatric Surgery* (3 ed., pp. 473-492). Philadelphia, Pa.; London: Saunders.
- Pless,I.B., & Nolan,T. (1991). Revision, replication and neglect--research on maladjustment in chronic illness. *J Child Psychol Psychiatry*, 32(2), 347-365.
- Polit,D.F., & Beck,C.T. (2004). *Nursing research: principles and methods* (7. ed.). Philadelphia: Lippincott Williams & Wilkins.
- Polit,D.F., & Hungler,B.P. (1999). *Nursing research: principles and methods* (6. ed.). Philadelphia: Lippincott Williams & Wilkins.
- Polit-O'Hara,D., Hungler,B.P., & Polit,D.F. (1997). *Essentials of nursing research: methods, appraisal, and utilization* (4. ed.). Philadelphia, Pa.: Lippincott-Raven.
- Rebok,G., Riley,A., Forrest,C., Starfield,B., Green,B., Robertson,J., et al. (2001). Elementary school-aged children's reports of their health: a cognitive interviewing study. *Qual Life Res*, 10(1), 59-70.
- Rintala,R. J., & Lindahl,H. (1995). Is normal bowel function possible after repair of intermediate and high anorectal malformations? *J Pediatr Surg*, 30(3), 491-494.
- Rintala,R.J., & Lindahl,H.G. (2001). Fecal continence in patients having undergone posterior sagittal anorectoplasty procedure for a high anorectal malformation improves at adolescence, as constipation disappears. *J Pediatr Surg*, 36(8), 1218-1221.
- Roach,M.A., Orsmond,G.I., & Barratt,M.S. (1999). Mothers and fathers of children with Down syndrome: parental stress and involvement in childcare. *Am J Ment Retard*, 104(5), 422-436.
- Sandstrom,M.J., & Schanberg,L.E. (2004). Peer rejection, social behavior, and psychological adjustment in children with juvenile rheumatic disease. *J Pediatr Psychol*, 29(1), 29-34.
- Sarkadi,A., & Bremberg,S. (2005). Socially unbiased parenting support on the Internet: a cross-sectional study of users of a large Swedish parenting website. *Child Care Health Dev*, 31(1), 43-52.
- Schmidt,S., Petersen,C., & Bullinger,M. (2003). Coping with chronic disease from the perspective of children and adolescents--a conceptual framework and its implications for participation. *Child Care Health Dev*, 29(1), 63-75.
- Silverman,D. (2001). *Interpreting qualitative data: methods for analyzing talk, text and interaction* (2. ed.). London: SAGE.
- Simons,J. (2002). Parents' support and satisfaction with their child's postoperative care. *Br J Nurs*, 11(22), 1442-1444, 1446-1449.

- Sobo,E.J. (2004). Pediatric nurses may misjudge parent communication preferences. *J Nurs Care Qual*, 19(3), 253-262.
- Stephens,F.D., & Durham Smith,E. (1986). Classification, identification, and assessment of surgical treatment of anorectal anomalies. Report of a work meeting held on May 25-27, 1984, at "wingspread" Convention Center, The Johnson Foundation, Racine, Wisconsin, USA. *Pediatric Surgery International*, 1(4), 200-205.
- Stephens,F.D., & Smith,E.D. (1971). *Ano-rectal malformations in children*. Chicago.
- Streisand,R., Swift,E., Wickmark,T., Chen,R., & Holmes,C.S. (2005). Pediatric parenting stress among parents of children with type 1 diabetes: the role of self-efficacy, responsibility, and fear. *J Pediatr Psychol*, 30(6), 513-521.
- Torrance,G.W., Feeny,D., & Furlong,W. (2001). Visual analog scales: do they have a role in the measurement of preferences for health states? *Med Decis Making*, 21(4), 329-334.
- Trajanovska,M., & Catto-Smith,A.G. (2005). Quality of life measures for fecal incontinence and their use in children. *J Gastroenterol Hepatol*, 20(6), 919-928.
- Tsuji,H., Okada,A., Nakai,H., Azuma,T., Yagi,M., & Kubota,A. (2002). Follow-up studies of anorectal malformations after posterior sagittal anorectoplasty. *J Pediatr Surg*, 37(11), 1529-1533.
- Uzark,K., & Jones,K. (2003). Parenting stress and children with heart disease. *J Pediatr Health Care*, 17(4), 163-168.
- van Kuyk,E.M., Wissink-Essink,M., Brugman-Boezeman,A.T., Oerlemans,H.M., Nijhuis-van der Sanden, M.W., Severijnen, R.S., et al. (2001). Multidisciplinary behavioral treatment of defecation problems: a controlled study in children with anorectal malformations. *J Pediatr Surg*, 36(9), 1350-1356.
- van Laerhoven,H., van der Zaag-Loonen,H.J., & Derkx,B.H. (2004). A comparison of Likert scale and visual analogue scales as response options in children's questionnaires. *Acta Paediatr*, 93(6), 830-835.
- Warschburger,P., Buchholz,H.T., & Petermann,F. (2004). Psychological adjustment in parents of young children with atopic dermatitis: which factors predict parental quality of life? *Br J Dermatol*, 150(2), 304-311.
- Witt,W.P., Riley,A.W., & Coiro,M.J. (2003). Childhood functional status, family stressors, and psychosocial adjustment among school-aged children with disabilities in the United States. *Arch Pediatr Adolesc Med*, 157(7), 687-695.
- Young,M.H., McMenamy,J.M., & Perrin,E.C. (2001). Parent advisory groups in pediatric practices: parents' and professionals' perceptions. *Arch Pediatr Adolesc Med*, 155(6), 692-698.
- Youngblut,J.M., Brooten,D., & Kuluz,J. (2005). Parents' reactions at 24-48 hrs after a preschool child's head injury. *Pediatr Crit Care Med*, 6(5), 550-556.