Neurogenic bladder and bowel dysfunction

- incontinence and life situation in adolescents and adults with spina bifida

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To all the fantastic children and families who I have had the privilege of meeting and getting to know and cooperate with in my role as a pediatric nurse and urotherapist.

Neurogenic bladder and bowel dysfunction - incontinence and life situation in adolescents and adults with spina bifida

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ABSTRACT

BACKGROUND: In individuals with myelomeningocele (MMC), survival rate has significantly increased over the past 40-50 years due to medical innovations. In the coming years, many adolescents will be transferred to adult care.

AIMS: To investigate life situation, quality of life, follow-up in adult care and the incidence and experience of incontinence in adults with MMC. To compare the incidence of incontinence with a youth group and investigate whether continence is one of the prerequisites for an active life and close intimate relationships.

METHODS AND RESULTS: **Study I** Sixty-nine adults (27–50 years) with MMC participated in a structured telephone interview. Pads for urinary and fecal incontinence were used by 87% and 14% had contact with a urotherapist. About 60% were single, 90% had attended high school and 67% had a job. **Study II** The validated HRQoL instrument SF-36 was answered by 61 of 69 individuals. There were significantly lower scores for the overall physical quality of life while scores for the overall mental quality of life were higher than for the reference group. Neither physical nor mental quality of life was affected by whether the individual had fecal incontinence, lived with a partner or had children. **Study IV** In a descriptive qualitative semi-structured interview, 9 adults described their personal experience of living with incontinence. **Study III** All 16-18-year old's with MMC (25) from western Sweden were included in a prospective cross-sectional study regarding urinary and fecal incontinence as well of life situation. All were followed according to the national care program and 68% (17/25) were urinary continent. Of these, 12 had an active social life and 8 had experience of having a partner. Of the 8 with incontinence, none had an active social life or a close physical intimate relationship.

CONCLUSION: Incontinence is common in adults with MMC and few have urotherapy support. Life situation and incontinence are not reflected in the generic HRQoL instrument SF-36. The adults' experience of how incontinence affects life is consistent with reports from individuals with acquired incontinence. In the adolescent study, the results indicate the importance of follow-up and active treatment strategies to achieve urinary continence. Continence seems to be a success factor for an active social life and close intimate relationships.

KEYWORDS: spina bifida, myelomeningocele, adults, life situation, follow-up, HRQoL adolescents, continence, incontinence

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SAMMANFATTNING PÅ SVENSKA

Hos vuxna med ryggmärgsbråck har överlevnaden ökat signifikant under de senaste 40–50 åren på grund av medicinska innovationer som t.ex. förbättrade operationsmetoder vid hydrocefalus och bevarande av njurfunktionen med hjälp av Ren Intermittent Kateterisering (RIK). Många ungdomar överförs därför nu till vuxenvården med ett stort behov av livslång vård och uppföljning av ett multiprofessionellt team. Urin- och avföringsinkontinens är mycket vanligt men ses ofta inte som orsak till behandling eller uppföljning. Syftet med avhandlingen var att hos vuxna individer med ryggmärgsbråck undersöka livssituationen, hälsorelaterad livskvalitet, förekomst av inkontinens, urologisk och uroterapeutisk uppföljning och upplevelsen av att leva med urininkontinens. Syftet var också att jämföra förekomst av inkontinens mellan vuxna och en ungdomsgrupp i åldern 16–18 år och i den senare även undersöka om kontinens är en av förutsättningarna för ett aktivt liv och nära intima relationer.

Studie I Sextionio individer (27–50 år) med ryggmärgsbråck deltog i en strukturerad telefonintervju. Av deltagarna använde 87% skydd för urin och/eller avföringsläckage och 14% hade kontakt med uroterapeut, drygt 60% var ensamstående, 90% hade gått på gymnasiet och 67% hade arbete. Av de individer i studien som inte genomgått någon urologisk operation följdes 69% sporadiskt eller inte alls inom vuxenvården.

Studie II Det validerade HRQoL-instrumentet SF-36 besvarades av 61 av 69 individer. Resultaten visade att studiegruppen hade signifikant lägre poäng i fysisk funktion, allmän hälsa och övergripande fysisk livskvalitet jämfört med referensgruppen. Den övergripande livskvaliteten för psykisk hälsa var högre än för referensgruppen. Varken fysisk eller psykisk livskvalitet påverkades av om individen hade avföringsinkontinens, partner eller barn.

Studie IV I en beskrivande kvalitativ semistrukturerad intervju berättar 9 individer från samma kohort som i studie I och II om sina personliga upplevelser av att leva med inkontinens.

Studie III Alla 16–18-åringar med ryggmärgsbråck (25 individer) från västra Sverige ingick i en prospektiv tvärsnittsstudie angående urin- och avföringsinkontinens samt deras livssituation. Alla följdes enligt ett nationellt vårdprogram. Av de 17 ungdomar (68%) som var urinkontinenta hade 12 ett aktivt socialt liv och 8 hade erfarenhet av att ha en partner. Av de 8 ungdomarna med urininkontinens hade ingen ett aktivt socialt liv, partner eller en nära intim relation.

Sammanfattningsvis tyder studierna på att inkontinens är vanligt förekommande hos vuxna med ryggmärgsbråck och att få har stöd av uroterapeut. Livssituation och inkontinens återspeglas inte i det generiska HRQoL instrumentet SF-36. De vuxnas upplevelse om hur inkontinens påverkar livet överensstämmer med rapporter från individer med förvärvad inkontinens. Individer med medfödd inkontinens verkar behöva samma tillgång till behandling som individer med förvärvad inkontinens. I tonårsstudien indikerar resultaten på vikten av uppföljning och aktiva behandlingsstrategier för att uppnå urinkontinens. Kontinens verkar vara en framgångsfaktor för ett aktivt socialt liv och nära intima relationer.

Nyckelord: ryggmärgsbråck, vuxna, ungdomar, livssituation, livskvalitet, uppföljning, kontinens, inkontinens

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LIST OF PAPERS

This thesis is based on the following studies, referred to in the text by their Roman numerals.

I. Vu Minh Arnell M, Seljee Svedberg K, Lindehall B, Jodal U, Abrahamsson K. Adults with myelomeningocele: An interview study about life situation and bladder and bowel management. J Pediatr Urol. 2013 Jun;9(3):267-71.

II. Vu Minh Arnell M, Seljee Svedberg K, Lindehall B, Möller A, Abrahamsson K. Health-related quality of life compared to life situation and incontinence in adults with myelomeningocele: Is SF-36 a reliable tool? J Pediatr Urol. 2013 Oct;9(5):559-66.

III. Vu Minh Arnell M, Abrahamsson K. Urinary continence appears to enhance social participation and intimate relations in adolescents with myelomeningocele. J Pediatr Urol. 2019 Feb;15(1):33. e1-33. e6.

IV. Vu Minh Arnell M, Korsgaard R, Abrahamsson K. The voice of adults with myelomeningocele - experience of urinary incontinence and how it affects life. In manuscript.

CONTENT

ABBREVIATIONS

Ch	Charrière catheter size, corresponds to millimeters in circumference
CIC	Clean Intermittent Catheterization
FCC	Family Centered Care
HRQoL	Health Related Quality of Life
MACE	Malone Antegrade Continence Enema
MCS	Mental component summary
MMC	Myelo Meningo Cele
NTD	Neural tube defects
PCS	Physical component summary
SF-36	The Short Form (36) Health Survey

DEFINITIONS IN SHORT

Ability to walk	Ambulatory everywhere or just walking indoor
Ambulatory ability	Walking without the need of a wheelchair
Assistance	Practical help
Bodily pain	Pain magnitude
Clean Intermittent Catheterization	The bladder is emptied regularly 4-6 times a day, with a clean catheter
Dysphoria	Interviewees is weeping during the interview or by the individual stating that he/she is depressed
Emotional role	Cut down time, accomplished less
Fecal continence	Dryness between regimens not including dripping of enema solution in a period up to 2 h after the enema was performed
General health	Sick easier, excellent health
Independence in CIC and fecal elimination regimens	Physically performing the total procedure without need of assistance
Intimate relations	Intimacy without sexual intercourse
Mental health	Nervous, happy, peaceful
Micturition chart	Volume of urine at each CIC and fluid were measured for 2 days
Participation in social life	At least twice a week spending free time with friends and being able to stay overnight when desired, without family or assistant attending.
Partner	Person with whom the individual can have intimate relations and/or sexual intercourse

Physical functioning	Mobility
Physical role	Limitations caused by physical health problems
Reminder	Help to remember the time for CIC
Sexual debut	First occasion of sexual intercourse
Social functioning	Social excellent, social time
Urinary continence	Totally dry during day and night without need of incontinence pads
Vitality	Energy, tiered
Young age	20-29 years of age

THESIS AT A GLANCE

	AIM	DESIGN/METHOD	RESULTS	CONCLUSION
I	To asses life situation, bladder and bowel management in adults with MMC after transferal to adult care.	Cross-sectional cohort study with 69 adults participated in an individual structured interview.	Of the individuals 90% had passed high school or had university education. Fifty-three (77%) had their own apartment. CIC was used by 71%. Of those with no urological operation, 31% had a consultation with urologist every 3 years. The corresponding number for those operated on were 53%.	Few of the participants had urotherapy support. The majority used pads, and none had support with their fecal elimination regimen. If pads were used it was harder to get employed. About 60% were single.
II	To investigate if life situation and incontinence in adults with MMC are reflected in HRQoL.	Cross-sectional, cohort study with 61of 69 from study I who answered the generic Health related Quality of life instrument SF-36.	The individuals had lower scores in the overall physical quality of life but higher scores in the overall mental quality of life.	Neither physical nor mental quality of life was affected by whether the individual had fecal incontinence, lived with a partner or had children.
III	To evaluate urinary continence in adolescents with MMC, involved in a urotherapy/urology program and if urinary continence is one condition to enable social participation and close physical intimacy.	Cross-sectional, quantitative, cohort study. A structured individual interview with 25 adolescents, were conducted. Prospective investigation about incontinence was implemented.	Seventeen of 25 (68%) were urinary continent. Of these, 12 had an active social life and 8 had experience of having a partner. Eight individuals were urinary incontinent. None of them had an active social life or a close physical intimate relationship.	The importance of follow-up and active treatment strategies to achieve urinary continence seem to be one of the predictors for having an active social life and the possibility of close physical intimate relationship.
IV	To describe experience of urinary incontinence and how incontinence affects life.	Cross-sectional, descriptive qualitative semi structured interview with 9 individuals from the cohort in study I and II.	The participants described similar experiences as individuals with acquired urinary incontinence.	Individuals with congenital incontinence seem to need the same access to treatment as individuals with acquired urinary incontinence.

1 INTRODUCTION

Spina bifida is a diagnosis within neural tube defects, (NTD). Globally about 300,000 children are annually born with NTD, and the most common malformation is myelomeningocele (MMC) (1). The individuals in this thesis have either MMC or Lipo-MMC.

Since the 1970s, there is a multi-professional team in Gothenburg that follows children and adolescents with MMC. The team at Queen Silvia Children´s hospital consists of urologist, pediatric urotherapist, neurologist, intestinal surgeon, occupational therapist and habilitation assistant. Our catchment area consists of western Sweden with a population of 2.3 million of the total 10,3 million inhabitants in Sweden 2019 (2). In the beginning of the 70s the incidence of MMC in Sweden was 5.5 per 10,000 births. In year 2016 the figure has decreased to 2,4 individuals in 10,000 births (3). The fact that MMC has become a rare diagnosis, the number of patients decreases in pediatric care while it still increases in adult care. During the last decades the pediatric MMC-team often discusses in terms of; - What happened to the individuals with MMC after leaving pediatric care and what does their life look like? These questions became the basis for my PhD-studies.

MYELOMENINGOCELE, MMC

Dysmorphology on the neural tube can lead to many different malformations known as, neural tube defects, (NTD). The malformations vary in extent and symptomatology. Major defects include anencephaly, a large open defect meaning survival is impossible. Open spina bifida, myelomeningocele (MMC), could be extensive with severe neurological injuries. On the other hand, occult spina bifida could just be a small defect in the vertebral arches, with normal neurological function (4,5). MMC is a malformation due to a lack of closure of the spinal canal and it occurs during the third to fourth week of pregnancy. The spinal cord is often deformed, and the nerve fibers are stretched and damaged. The cause of myelomeningocele is not completely clarified (4). Nowadays, in the industrial world, there is a decline in the numbers of children born with MMC. There is a consensus that if the mother takes folic acid or eats food fortified with folic acid the risk of NTDs in the fetus declines (5,6). Another reason for the decline may be the increased number of abortions following prenatal screening (3). Certain heredity, and probably environmental factors can also affect the occurrence of NTD (5). During the last 60 years in Sweden, care for individuals with MMC has developed. Due to the improved hygiene conditions at Swedish hospitals during the 1950s, newborns with MMC could survive. Neurosurgeons began to operate on and cover the hernia in plastic surgery procedures, and in late 1950s, the first shunt procedure was carried out on at a little boy in the US. The first shunt operation was carried out on an individual with MMC in Sweden in the early 1960s. To be able to close the myelomeningocele and to do a shunt operation for hydrocephalus were two important conditions for survival (7). Today, most children with the condition survive, and we can expect most of them to reach adulthood. However, we do not really know how aging will affect individuals with MMC. There are articles that claim that adults with MMC have more hospital stays than the general population, they have more problems with pain, obesity, pressure ulcers, high blood pressure, and even premature death from uremia (8-11).

NORMAL BLADDER FUNCTION

At normal function the bladder stores the urine under low pressure and without leakage. Individuals should be able to decide voluntarily when to empty the bladder and should be able to empty it completely. Getting the micturition cycle to work depends on a functioning nervous system. When the bladder is filled during the storage phase, the sympathetic branch of the autonomic nervous system ensures that the detrusor is relaxed, and the internal sphincter is contracted. Voluntary micturition starts at brain level, the parasympathetic branch of the autonomic nervous system giving opposite signals, the detrusor contracting and the sphincter relaxing. The flow continues until the bladder is emptied. The detrusor muscle of the bladder consists of smooth muscle that ends in the bladder neck area where the inner sphincter is located. The urethra consists of both smooth and striated muscles. The striated muscles form the outer sphincter are will-controlled. Good pelvic floor muscles are important to counteract rapid rises of pressure in the abdominal cavity and to maintain urinary continence (12,13).

NEUROGENIC BLADDER DYSFUNCTION

Neurogenic bladder dysfunction in MMC, almost always means a loss of function. Normal function depends on the nervous control from the brain via nerve roots in the spinal cord and peripheral nerves to the bladder and urethral muscles being intact (13). In neurogenic bladder dysfunction the nerves, which are important for emptying the bladder are damaged. The emptying of the bladder and the relaxation of the urethra sphincter which are usually coordinated, are dyssynergic. Neurogenic bladder dysfunction in individuals with MMC entails both a peripheral nerve and spinal cord damage. It is not usually possible to make a diagnosis according to the level of the location of the MMC, so the diagnosis has to be made according to a careful mapping of the function. About 10% of individuals have a normal bladder function while 90% have one of the four types of neurogenic bladder dysfunction below. (14).

	Sphincter +	**Sphincter** -
Detrusor +	~35% High-risk bladder	~10% Low-risk bladder
Detrusor -	~10% Risk-bladder	~35% Low-risk bladder

NORMAL BOWEL FUNCTION

During bowel movements, an interaction between the colon, rectum and sphincter muscles begins. For most individuals, the urge to empty the bowel starts after eating. It is of importance to take advantage of the gastrocolic reflex. The feces in the colon are transported down into the rectum by a strong contraction. When the wall in the rectum expands, signals go through the pelvic nerve and a feeling of urgency arises. The bowel movement is started by influencing the pelvic floor, which then relaxes, and the anorectal angle is straightened. Usually a short straining is needed to start the emptying, which takes place automatically (15).

NEUROGENIC BOWEL DYSFUNCTION

About 90% of the individuals with MMC have a neurogenic bowel dysfunction (16). Due to the congenital damage of the spinal cord, the intestine is affected with a lack of emptying reflex and there is also slower peristalsis to the colon and a weak rectal sphincter. These factors lead to both constipation and fecal incontinence in this patient group (17,18).

Bowel regimens

Experience gained at our pediatric MMC-clinic since the 1970s, tells us that most of our patients need some kind of bowel emptying regimens. Every child gets an individual treatment plan according to the national and local care program (19). As soon as the child can sit up on a potty chair or on a special toilet seat, we initiate bowel regimen while sitting. The goals are to sit on a toilet when having an enema, that all feces should get into the toilet and that there will be no fecal leakage in between the times of the enemas. The definition “to be without leakage in between the times of the enemas” is sometimes called pseudo-continence by Vande Velde et al (20,21). The method must be effective and sufficiently simple for patients to perform so they are able to handle the procedure independently as they get older. A review from Belgium establishes the importance of an individually stepwise adapted method for bowel emptying and concludes that conventional methods must be well-proven before any surgical procedure is considered. The importance of lifelong follow-up of the emptying routine is emphasized for all individuals with MMC (21). In a registry study from the United States, the bowel emptying methods were compared for three groups; children, adolescents and adults with MMC. Adults more often used digital stimulation, and colostomies were not uncommon. Only 17% of the adults used enemas compared with 27% in children (22). On the market, there is a variety of tools to administer an enema. For individuals with MMC the principle is to get the enema fluid to remain in the bowel for long enough to mimic the emptying reflex. In a retrospective study of individuals with MMC from 2 to 24 years of age, and who followed a bowel-emptying program, Schletker et al found that the most common challenge when administering an enema was leakage of the solution during infusion (23). Being able to follow bowel regimens independently is important for reasons of integrity and self-esteem. However, in children up to 16 years of age it is hard to administer an enema independently, either trans-anal or antegrade like in Malone Antegrade Continence Enema (MACE) (24). In a study from the UK, parents found it hard to hand over responsibility to their teenagers and teenagers found it difficult to become independent in the enema regimen (25).

UROTHERAPY

The first university education in urotherapy in the world started in 1987 at the University of Gothenburg. To become a urotherapist you must have a university degree and have qualifications including registered nurse, physiotherapist or physician. To be able to apply for the urotherapy course nurses also need a specialization degree (26). “Urotherapy” is a combination of the word “uro” - from urology that means study of treatment and diseases of the urogenital tract and “therapy” - which means treatment of diseases or disorders by rehabilitation or curative process” a quote from UTF-Nordic (Urotherapeutic association-Nordic) (on internet) (27). A urotherapist investigates, treats and makes follow-up interventions to deal with both bladder and bowel problems including leakage, urgency and emptying difficulties. The urotherapist conveys knowledge and understanding of cause and symptoms of the condition in order to achieve the best treatment results. The urotherapist have also education and knowledge about prescription of products for urinary and fecal incontinence and retention (26).

Important conditions for survival in this patient group were surgical closure of the myelomeningocele in the 1950s and shunting of hydrocephalus in the 1960s. However, there was still one major problem that affected basically all individuals with MMC, namely the inability to empty the bladder which entailed a high risk of kidney deterioration, resulting in uremia with a fatal outcome. In the late 1970s, Clean Intermittent Catheterization (CIC) was introduced in Sweden and it became a revolution for individuals with MMC. From having a high risk of renal failure, the risk was now considerably minimized. The method was first described by Lapides et al in 1972 and introduced in Sweden in 1977 by Lindehall and Hellström. Regular complete emptying with a clean catheter was more important than a sterile technique (28,29).

Before CIC was introduced, parents of children with MMC learned manual compression of the child's bladder and later children had to try abdominal straining in order to empty the bladder. These methods increased the risks of vesicoureteral reflux (29). In the medical world, a non-sterile method of catheterization was not a possibility. However, Lapides showed that two things were more important than a sterile catheter, a regular and an effective emptying with bladder volumes for an adult under 400 ml. The bacteria were inevitably introduced into the bladder but drained again after 3-4-hour intervals daytime. The CIC method became the second-best method for bladder emptying (28,30).

As the majority of children with MMC have neurogenic bladder dysfunction, CIC is vital and lifelong (31). From a medical point of view, the bladder emptying is the most serious problem. It means that when a child with MMC is newborn, CIC should be introduced as soon as possible. This is gold standard in many of the industrial countries (32). The urotherapist's most important role in the natal period is to teach and support the parents in how to perform CIC on their child. A child with MMC will be followed at the outpatient clinic in Queen Silvia Children`s hospital for 18 years. The team is inspired by “family-centered care” FCC, a way to work with and for the family and their child with special needs. Parents and professionals should be seen as partners where continuity in care and accessibility are two key concepts. Respect, family strength and collaboration are central in FCC (33). In our unit, each child has a personal urotherapist who follows the child and knows the family.

In an outpatient clinic for children and adolescents with MMC, most of the patients are expected to have the diagnosis neurogenic bladder and bowel dysfunction. The urotherapist has a prior knowledge of the diagnosis and a structured national and local care program to follow (20). Urodynamic examinations play a central role in the investigation and follow-up of neurogenic bladder dysfunction and are performed by the child´s personal urotherapist (34-37). An important complement to urodynamics is observation of the micturition list, leakage test and registration of bowel emptying. The urotherapist is responsible for the child receiving individually tested pads or diapers for leakage and catheters for CIC. Changes in the micturition or continence pattern should lead to follow-up cystometry. In connection with the visit an ultrasound is made after CIC or micturition to check that the child empties the bladder completely. In neonates and infants, a 4-hour micturition observation with provocation and ultrasound after micturition are conducted (38,39).

From the integrity aspect, learning self CIC is of great importance. Most children learn to perform CIC by an age of 6-9 years old. However, the urothearapist and the parents may encourage the child to practice as soon as the child shows interest in any part of the treatment. Depending on the child's conditions such as hand function and cognitive ability, CIC is practiced in small steps and always adapted individually. If the child has a difficulty in time perception, the CIC may be related to another event such as meals (40-42). The urotherapist starts early to motivate the child and the family to introduce self-CIC. We invite the child and parents to “CIC school” where they can meet others in the same age who also come to practice self-CIC.

LIFE SITUATION

During the last 40 years, individuals with MMC survive into adulthood. During these years it has been noticed that it is not only the physical problems that are in focus. Together with the cognitive problems, the entire life situation is affected. Almost 20 years ago, in 2001, an article was published by Bowman et al in which they pointed out that a major challenge was providing care for the growing adult population. Such care entails multidisciplinary follow-up for patients and an active network of healthcare professionals in adult care (43). Today, there is knowledge about what affects the life situation in individuals with MMC. Urine and fecal leakage are major problems and can be a contributing factor to loneliness (44). A life without social context like having a job, a partner or friends, affects health and leads to difficulties in life conditions. The physical problems become more and more accentuated the older you get. Inactivity leads to pain, obesity and incontinence which can contribute to pressure ulcers (45,46). In a study from the US they look at the fact that adults with MMC have physical and cognitive problems that affect their psychosocial situations which in turn can lead to mental illness. Since 2018, the Spina Bifida Association in the US has provided; Guidelines for the Care of People with Spina Bifida, with the intention of helping individuals achieve good mental health throughout life (47). Not being active in the society can contribute to loneliness and affect life situation. Barf et al found that young adults with MMC experienced difficulties in participating in the society due to physical or mental limitations (48). In a study on friendship, young people with MMC often described a relationship with a particular friend as close and called this friend their best friend. However, this did not correspond to the friend's opinion. The conclusion was that young people with MMC experience differences in the quality and reciprocity of friendship compared with their peers (49). Still 20 years after Bowman's article, we face the same challenge.

COGNITIVE FUNCTION

Over the years, meeting many children and adolescents with MMC, we have noticed similar features among the individuals that we initially did not fully understand. However, many parents had the same experience. Their children had e.g. difficulties in mathematics, problems with memory, to interact with other children, and they had to be constantly pushed to get something done.

In the 21 centuries, research has shown that most individuals with MMC have cognitive difficulties due to primary brain dysmorphology and the occurrence of hydrocephalus (50-52). The published articles have pointed out how this affects life situation for both children and adults (53-55). A Swedish study showed that the individuals have difficulties in estimating time, understanding time perspectives and plan time (54). Peny-Dahlstrand et al described in an article the executive difficulties in children with MMC. The hardest thing is not to learn how to do things but to get things done (53). This sentence says a lot about the challenge that is faced by the individual, the family and by the professionals. In a urotherapeutic unit where we meet individuals with MMC, we start CIC and bowel emptying regimens where the goal is to create independence in vital and lifelong treatments. If CIC is not performed, even if the person knows how, it could become a life-threatening situation resulting in renal failure (55). Aware of the cognitive and executive difficulties affecting individuals with MMC, it is of great importance to have an, of the diagnosis experienced occupational therapist, in the team.

2 AIM

Study I
To assess life situation, bladder and bowel management and urological follow-up in individuals with MMC after transferal to adult medical care.

Study II
To evaluate HRQoL in adults with MMC measured by SF-36.

Study III
To evaluate urinary continence in adolescents with MMC, who are actively involved in a urotherapy/urology program. To evaluate if urinary continence is one of the conditions required to enable social participation and close physical intimacy.

Study IV
To describe the experience of urinary incontinence in individuals with MMC and how incontinence affects their lives.

3 PARTICIPANTS AND METHODS

Table 1. Overview of methodological approaches.

STUDY	**STUDY POPULATION**	**STUDY DESIGN**	**DATA COLLECTION**	**DATA ANALYSIS**
I	69 participants 37 females Age Md 34 yrs (27-50)	Observational, Cross-sectional, Quantitative, cohort study	Individual structured interview	Fisher´s exact test, p<0.05
II	61 participants 30 females Age Md 34 yrs (27-50)	Observational, Cross-sectional, Quantitative, cohort study	Questionnaire study	Descriptive statistics, Fisher´s exact test, Mann-Whitney´s test, Kruskal-Wallis´ test, p<0.05
III	25 participants 10 females Age Md 17 yrs (16-18)	Observational, Cross-sectional, Quantitative, cohort study	Individual structured interview. Prospective investigations	Descriptive statistics
IV	9 participants 7 females Age Md 46 yrs (40-56)	Observational, Cross-sectional, Descriptive Qualitative study	Individual semi-structured interview. Selection of participants was purposeful	Qualitative Content Analysis

STUDY POPULATION (Fig)

All children newborn to 18 years of age, with MMC and neurogenic bladder dysfunction, living in western Sweden, and attending Regional Rehabilitation Centre for Children and Adolescents. Inclusion criteria for the study population in study I, II and IV were patients with MMC, born before 1981. As children they had lived in western Sweden and had been assessed on at least two occasions by a pediatric urologist at Regional Rehabilitation Center for Children and Adolescents. They had been transferred to adult medical care before 2001.

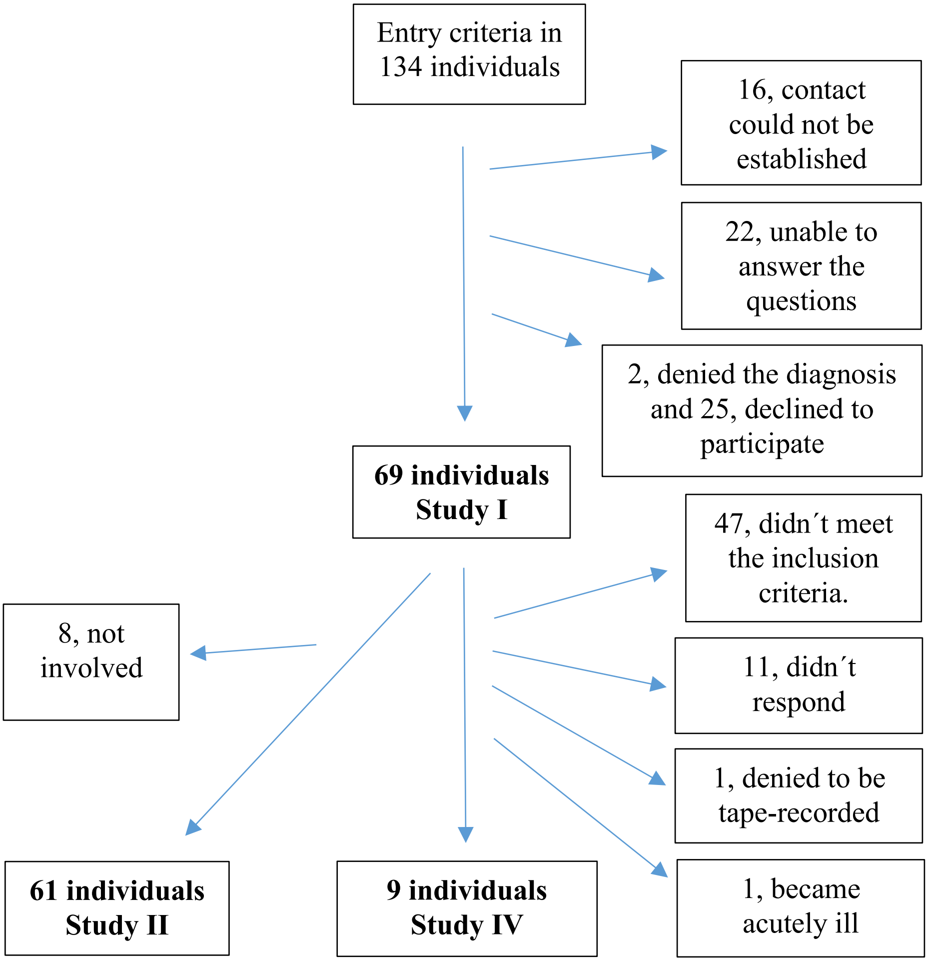

Figure. Individuals who met the inclusion criteria in study I, II and IV and drop-outs.

In **study I** 69 individuals (58%) participated. Twelve participants did not have a shunt due to hydrocephalus. Wheelchair was never used by 18, occasionally by 11, and always used by 40 participants. No evident differences in regard to age, gender, type of spina bifida, lesion level and having a shunt for hydrocephalus were noted between the group that declined to participate and the group that agreed. On the same occasion as interviews for study I took place, 61 of the 69 individuals answered the Heath Related Quality of Life questionnaire, SF-36, **study II.** Eight individuals participated in the pilot study for study I and did therefore not take part in study II. Four individuals only partly completed SF-36 because two became too emotional and two did not understand the questions. The study's selection of informants in **study IV** was purposeful for the aim and the method used. We intended to interview individuals from study I who had urinary incontinence and who, during the interview in study I, openly expressed opinions about their incontinence. The day before the planned interview one individual became acutely ill and one was anxious about being tape-recorded. All but one in study IV had a shunt.

Study III. All 25 individuals with MMC from the ages of 16 to18 years, living in western Sweden, and born between 1996 and 1998, agreed to participate in a face-to-face interview at the same occasion as the yearly urological/uroterapeutic follow-up. The interviews were performed at the outpatient clinic.

STUDY DESIGN

All studies were observational and cross-sectional. Study I, II and III were also quantitative as well as cohort studies. The cohort was defined as all individuals who met the inclusion criteria for participating in the studies. In study IV the design was a descriptive qualitative semi-structured interview. The participants were selected from study I. With purposeful selection means that you want to get the best and as much information as possible about the topic. The individuals were selected for the research project based on their knowledge and experience (56).

DATA COLLECTION

Study I

During 2007-2008, two pediatric urotherapists, who had never been involved in the care of the study individuals, conducted a structured telephone interview according to a protocol. The interview questions focused on the individuals´ life situation and bladder and bowel function. The personal opinion of the individuals characterized the answers. The mean time for the interviews was 58 min (range 30-120). As control material Official Statistics of Sweden data was used (57).

Study II

SF-36 was used as a generic HRQoL protocol evaluating eight domains: physical functioning (PF), role physical (limitation caused by physical health problems) (RP), bodily pain (BP), general health (GH), vitality (VT), social functioning (SF), role emotional (limitation caused by emotional problems) (RE), and mental health (MH) (58). The eight domains could be aggregated into two summary measures: physical component summary (PCS) and mental component summary (MCS). Scores were measured on a scale from 0 to 100, with 0 representing the lowest health status and with 100 representing the highest health status. Values for PCS < 20 or > 58 and MCS < 17 and >62 are calculated as artifacts (59). The questions asked during the telephone interview were done in accordance with the SF-36 Swedish manual for the interview user (58).

Study IV

The individuals were free to choose where the interview should take place but due to the nature of the issue, it had to be in a private room with no one else to overhear. The interviews were conducted in the participants´ homes or in a private room at the Regional Rehabilitation Center for Children and Adolescents. A pediatric nurse and urotheraphist, not involved in the care of the participants but with experience of the diagnosis, conducted the interviews. The interview was a tape-recorded face-to-face, semi-structured interview with two open-ended questions. All individuals were asked to share their experience of living with urinary incontinence and how the incontinence affected their lives. It was of great importance that the interviewer did not lead the interview by suggesting different predetermined areas, just be supportive by saying such as “Tell me more about this. Can you explain further?” The interviews lasted between 30 and 120 minutes.

Study III

During 2014, a urotherapist with knowledge of the participants conducted a structured interview with a questionnaire. All 25 individuals agreed to participate (100%). The questions asked were about urinary and fecal continence, CIC, bowel regimens, the need of reminder or help from assistant, social participation, physical intimacy with a partner and sexual debut. To have an objective observation, prospectively urinary continence was evaluated with 24-hour leakage test or a two-day micturition chart for those who reported that they were continent and did not have pads for incontinence prescribed. A 24-hour leakage test was performed with: pre-weighed pads, each in its own airtight plastic bag. The pads were changed every third hour (CIC interval) during daytime. The night pad was changed in the morning. After use, they were put back into the plastic bag which was sealed and returned to the hospital ward. Leakage was evaluated by weighing the pads and CIC volume and fluid intake was also measured.

DATA ANALYSIS

Descriptive statistics (Study II, III)

Continuous variables were expressed as mean, standard deviation (SD), median, min and max. Number and/or percentage described categorical variables.

Statistical analysis

For comparison between groups, a Mann-Whitney U test was used for continuous variables (study II) and Fischer´s exact test for dichotomous variables (study I, II). For tests between three or more groups with regard to continuous variables, a Kruskal-Wallis test was used (study II). All tests were two-tailed and conducted at a 5% statistical level. All statistical analyses were performed by using SAS_9.2 (Cary, NC, USA) or Statistical Package for Social Science (SPSS) for Windows 18.0.

Qualitative Content Analysis, QCA

As method used in study IV, qualitative content analysis, was conducted according to the technique described by Krippendorff (60). Data were analyzed with an inductive approach based on the content of the interview texts. To get a sense of the whole and identify what was relevant to the issue, the text was read several times. The text was then condensed to find the significant core or pattern. The analysis was described step by step to give the reader an idea of the thoughts of the individuals described in the content. Quotes from the interviews also exemplify the content. The first step was to find meaning units that answered to the research question. The meaning unit were condensed to shorten the text but retain the content, then the condensed meaning units were coded. To reflect the core of the interviews, categories were created, and categories were then divided into subcategories (61). Quotes from the participants from the interview material, to exemplify each subcategory, were illustrated in the text. The text was analyzed with a manifest qualitative content analysis, which means that no interpretation of the text was made.

ETHICAL CONSIDERATION

Study designs for study I, II and III were accepted by the Regional Ethical Review Board in Gothenburg and the study design for Study IV was approved by the Ethics Review Authority in Uppsala, Sweden. Participation was voluntary and informed consents were obtained. The ethical principles of the Declaration of Helsinki - Ethical Principles for Medical Research Involving Human Subjects - were followed. They included the principle stating the care of the individual must take precedence over the interests of both science and society (62). These principles were followed in the dissertation. In Study I, III and IV, where there were sensitive topics and questions of a private nature, we were careful to respect decisions of individuals not to answer. If participants in interviews raised issues related to healthcare contacts, we tried to help solve them. In connection with requests to participate in the studies, no more than two reminders were sent. Information was given to the participants that results would be treated confidentially.

4 RESULTS AND DISCUSSION

1. Life situation in adults

In a telephone interview study with 69 adult individuals 27-50 years of age, many expressed frustration and despair over their life situation. Many of the participants lived isolated lives far beyond what they had expected when young. As many as 90% of the individuals had gone to high school and over 50% had a driving license, but despite this several participants told that it was difficult to get a job, and difficult to keep it. Of participants, 67% had some form of employment, but 26% had no daily activity at all, corresponding to 7% in the control group from Official Statistics of Sweden (57). Many of the males pointed out that they lived alone (75%) and longed for someone to live with. Among females, about 50% lived with a partner. Internationally, Sweden stands out with 53 of 69 adult individuals with MMC having their own housing, but at the same time only 27 individuals of 69 told they were independent. This means that many of those living in their own homes need some sort of support. Several adults described how their parents, in their 80s, helped with cleaning and enemas (63). International studies have shown that many adults with MMC continue to live at home with their parent's (64-66). Asking questions about sexual experience, when the individual said he or she never had had a close intimate relation, felt unethical. What emerged was that out of those who now lived in a relationship, were also those who answered yes to the question on physically intimate relationships. Of the females 19% had children compared to 6% of the males. Corresponding figures from the Swedish population show that 86% of females and 82% of males had children (57).

Table 2. Life situation and civil status in 69 individuals (32 males and 37 females) with MMC. Control-group from Statistics Sweden 2011

		No. (%)	**Control-Group %**
Ambulatory		29 (42)	
Shunt		57 (83)	
Educational level			
	School for intellectual disability	5 (7)	
	Elementary school	2 (3)	
	High school	51 (74)	76
	University	11 (16)	23
Driving license			
	Yes	37 (54)	82
Employment			
	No daily activity	18 (26)	7
	Day center	3 (4)	
	Education	2 (3)	
	Employment	46 (67)	
Accommodation			
	Own living	53 (77)	
	Living in institution	10 (14)	
	Living with parents	6 (9)	
Independent			
	Yes	27 (39)	
	No	42 (61)	
Single	Total	42 (61)	
	Males	24 (75)	
	Females	18 (49)	
Sexual experience	Total	27 (39)	
	Males	8 (25)	
	Females	19 (51)	
Individuals with offspring	Total	9 (13)	
	Males	2 (6)	82
	Females	7 (19)	86

2. Urinary continence and bladder/bowel management

Table 3. Urine and fecal continence, independence and support; comparison between adults and teenagers with MMC.

Study I		**Study III**
Adults 27-50 yrs (no 69)		Teenagers 16-18 yrs (no 25)
29%	Urinary continence	68%
45%	Fecal continence	76%
13%	No pads	56
71%	CIC	100%
22%	Catheter size, 10 Ch	0
12%	Anticholinergic drugs	44%
94%	Independent in CIC	100%
14%	Urotherapy support	100%

When the results from study I were compared with study III, it turned out that adult individuals with MMC had not been given the opportunity to participate in the development of urotherapeutic care and bowel regimens. Many of the adults still had exactly the same sizes of pads, diapers or catheters that they had when they left pediatric care 10 to 30 years ago. Some pointed out that they had stopped CIC as it was too difficult to get catheters prescribed. One man told us, “If you get catheters prescribed there are just enough for weekends only, when I meet my friends, and the rest of the week I have to wear big diapers.” Today there are studies that show that a thicker catheter is a better choice than a thin one. There is a reduced risk for complication and the bladder empties faster, more efficiently and it is easier to empty completely (28,29). In study I, 22% of the adults used catheter ch10, a size suitable for preschool children with MMC. None of the teenagers used catheters ch10. CIC was conducted in 100% of teenagers and 71% of adults. Anticholinergics were used by 12% of adults, and 44% of adolescents. When comparing the continence levels of the two groups, there were large differences. In the youth group, 68% were continent for urine and 56% did not use pads at all. Comparative figures in the adult study showed a different situation where 29% were continent for urine and 13% did not use incontinence pads.

In adult care, it is expressed that healthy individuals cannot be followed up for preventive purposes. CIC or bowel regimens are not regarding as requiring follow-ups. Of the adults in the study, 14% had contact with a urotherapist but no one had contact regarding the bowel emptying regimens. In the youth group, 100% followed a national care program and all had support for bladder and bowel regimens. Since 2018 there is also a follow-up program for adults with MMC (19).

3. Social participation and intimacy in relation to continence

Table 4. Social participation and intimacy in 25 individuals with MMC between 16-18 years of age.

	25 participants	
Urinary continent no.17		Urinary incontinent no.8
12	Social participation	0
8	Physical intimacy	0
5	Sexual intercourse	0
11 (65%)	Males	4 (50%)
8 (47%)	Ambulatory (11)	3 (38%)
12 (71%)	Shunt (20)	8 (100%)

In study III, 25 teenagers from 16 to 18 years of age were interviewed about current lives. The results of the study showed that 17 out of 25 individuals were urinary continent. To reach continence, 9 had undergone continence surgery, 6 had CIC in combination with anticholinergic treatment and 2 were continent using CIC only. Eight teenagers were incontinent, 5 of those had tried anticholinergic treatment to enhance continence but had finished the medication as it did not give desired result. Another 3 used anticholinergics but were still incontinent due to sphincter insufficiency. All 8, had been offered continence surgery but had declined.

Of the individuals who were continent, 12 of 17 had a shunt and of those who were incontinent all had a shunt. We discovered that 12 of the 17 teenagers who were continent expressed that they were active in social life and had sports activities, met friends and were not afraid to sleep over at a friend's home. Eight of the 12 who were active in social life had experience of a close physical intimate relation. In those with urinary incontinent, no one said that they had a social life with activities outside the home, no one said they met friends in their free time, and no one had experience of close physical intimacy. In a qualitative study from Canada about incontinence and social participation, children and adolescents who were continent had more possibilities for social participation (67). However, we are aware that incontinence may not be the only reason why individuals with MMC do not socialize with friends. In a Palestinian study with children and youth with MMC it was shown more like a total stigmatization about their disability and not only their incontinence situation that made them lonely, vulnerable and without an active social life (68). Individuals cognitive difficulties can also contribute to exclusion (69). In a qualitative interview study (IV) where 9 adults with MMC described their experience of living with incontinence, the results showed that incontinence had a great impact on their lives. In our study we recognized that the individuals with MMC described similar experiences to those suffering from acquired incontinence. The participants described their thoughts about urinary leakage. The results are not generalizable as this was a qualitative study. However, the experience of congenital urinary incontinence and ways of adapting life is consistent with reports from participants with acquired incontinence (70-72).

4. Questionnaire and Interview

Table 5. HRQoL SF-36 and statistics. Participants versus Swedish population.

Domains	**Participants, mean**	**Referents, mean**	**p-value**
Physical function	42,2	93,0	<0.0001
Role physical	64,8	88,9	<0.0001
Body pain	75,2	76,9	ns
General health	67,7	79,1	<0.0001
Vitality	63,1	68,4	ns
Social function	82,2	88,5	ns
Emotional role	81,4	87,4	ns
Mental health	74,0	80,9	ns
Physical component summary	34,5	52,1	<0.0001
Mental component summary	51,0	49,4	<0.0001

Table 6. Life situation compared to HRQoL: PCS and MCS

Variable	**PCS**	**PCS,** **p-value**	**MCS**	**MCS,** **p-value**
Sex		ns		ns
Age, 20-29	increased	p<0.05		ns
Age, 30-50		ns		ns
Shunt		ns		ns
Walking, yes		ns	decreased	p<0.05
Children		ns		ns
Partnership		ns		ns
Employment, yes	increased	p<0.05		ns
University		ns		ns
Pos. experience		ns		ns
Neg. experience		ns	decreased	p<0.05
Dysphoria, yes	decreased	p<0.05		ns
Incontinence urine		ns		ns
Incontinence feces		ns		ns

In study I, several of the participants cried during the telephone interview and some expressed that they were dysphoric. They said that they felt forgotten and lonely. We also asked participants to answer SF-36, which is one of the world's most widely used generic instrument for measuring physical and mental health, study II. The questions were difficult for the individuals to answer, such as asking them how they felt now compared to four weeks ago, this due to the individuals' memory problems (51). We could also see that the scale with six different answer options were too difficult for participants, which is in accordance with the findings of Lindqvist and Dennis (73,74). Sometimes the individual did not answer what was intended by the question. The results showed, despite the individuals saying that they were dysphoric, that the overall mental health (MCS) was better than in the reference group. The total group had significantly lower scores, compared to the reference group in physical component summary (PCS). HRQoL, both physical and mental, was not affected by whether the individual had fecal incontinence, had a partner or had children. The PCS was lower but MCS was unaffected in the 12 individuals who expressed dysphoria during the interview. We considered that for individuals with MMC, SF-36 was not a reliable tool for measuring HRQoL. A better instrument to assess QoL in individuals with MMC is required.

5 GENERAL DISCUSSION AND FUTURE PERSPECTIVES

In this dissertation, the overall questions were life situation and the prevalence of incontinence in adults and adolescents with MMC. The quantitative interview studies focused on the life situation of adults, such as education, work, housing and personal partnership. Urine elimination, incontinence and urological and urotherapeutic follow-up in adult care were also evaluated. To investigate whether the adult individuals' HRQoL was affected by the life situation and the incidence of incontinence, they answered SF-36, a generic HRQoL form with a focus on physical and mental health. In the fourth article, a qualitative interview, with focus on the individual's personal experience of living with incontinence and the way it affected their lives. In the adolescent group, who are followed through a urological/urotherapeutic care program, we prospectively evaluated urinary incontinence. Two issues were central, whether urinary continence affects the ability to live an active life in society and whether urinary continence is one of the pre-conditions for close physical intimacy.

Life situation

We chose to interview adult individuals by telephone so they would have the opportunity to answer the questions themselves. Furthermore, the study is not an objective observation but based on the personal opinion, thus, the reality for the individual. It is not uncommon when postal questionnaires are to be answered, individuals with MMC often receive help from relatives or assistants (64). A telephone interview was also a conscious choice as dropouts are common when individuals have to remember the place and time for an interview as their prospective memory both for time and events are impaired (51,54,73,75). Some studies have analyzed success factors for having a job, such as male gender and the ability to walk (75,76). In our study, it turned out that the significant factors to get a job were university education and continence, not to male gender, level of independence or ambulatory status. Taking this into account, Sweden is very well adapted to the disabled (78). Törnbom et al followed a group of individuals with spina bifida and cerebral palsy regarding their work situation. When they compared the group of individuals in 1997 with 2009, the number of individuals who had a job decreased significantly, but the level of education was a success factor for having a job (79) which also emerged in our study.

Lifelong follow-up and support

Today, we have extensive knowledge in several aspects of MMC. The condition is often associated with a great need for care due to complex physical and cognitive profile (47,51,53,55). In pediatric care, we work preventively in the care of individuals with MMC. At the age of 18 and followed up in Sweden since birth, there are no individuals with severe kidney damage (19). In our adult study participants who had not undergone urological surgery 69% were in adult care, followed very sporadically or not at all. We are convinced that these adults were undertreated in comparison to the adolescents with the same conditions. By urologists in adult care, it is a common opinion that they cannot follow individuals without obvious medical needs. The ICCS (International Children's Continence Society) recommendation of follow-up in adult care is every third year with ultrasound over the urinary tract but at the same time be checked closer with regard to CIC routines and urinary tract infections (80). With improved pediatric care, it is now the adults who become ill. There should be no reason why adult individuals not could have healthy kidneys during their lifetime (81). A regular multi-professional and urological follow-up that involves preservation of kidney function, CIC checkup and medication to maintain continence must become real for all adults with MMC (81,82). Close cooperation between the urologist and the urotherapist is necessary for follow-up when it comes to CIC, bowel regimens, medical treatment and continence surgery. It is medically difficult for several reasons to achieve urinary continence in this patient group (81,90). To reach the best results medical professionals must build up a trusting and respectful relationship with patients, to lead and support them so they can cope with vital, lifelong treatments. The adults need a lot of help, which is time-consuming for healthcare professionals and it takes years of experience of the condition to understand the needs of individuals with MMC. In a study from Miller et al where parents of children with spina bifida expressed what was important to them in the care of their child. The desire of continuity in care, thorough knowledge of the child and its diagnosis, communication and telephone follow-up and available care coordinator were predominated (83). The same desire the parents had for their child and for themselves could be a good suggestion for how the care would be structured even for adult individuals with MMC.

CIC, incontinence and HRQoL

The individuals in study I told that they experienced low levels of competence in the primary healthcare professionals with respect to products for urine retention and incontinence. It was obvious that the persons neither were guided in updating their aids, nor in following the development of products or new research in the field. This shows that education and up-dated information are required for those who prescribe aids for CIC and incontinence.

There have been discussions among urologists that urinary incontinence in individuals with MMC does not appear to affect HRQoL negatively (84-86). Ehrén et al reported that only 20% of the adult individuals with MMC in their study used anticholinergic drugs, and one explanation could be that urinary leakage was not a major problem for the individual (87). When we evaluated the results of SF-36 in this patient group we found that life situation and incontinence were not truly reflected by this instrument. With a diagnosis specific HRQoL instrument, Szymanski et al have been able to show that children with MMC and urinary incontinence had impaired HRQoL already at the age of 10 (88). In addition, Verheof et al reported that 69% of young adults with MMC perceive their urinary incontinence as a problem and in a study from US the authors reported that HRQoL is impaired in adults with MMC who have urinary incontinence (88,89). In the adolescents' study we define continence as, "always dry" without prescription of incontinence pads, which correspond to the definition in a French study (90). The definition of urinary incontinence is not consistent in the literature with regard to individuals with MMC. The most common definition was "always dry" but only 37% of the authors described which method they had used to evaluate the continence status of participants (91). In another registry study from the National Spina Bifida Patient Registry in US "Less than once per month" were considered as continent (22). Furthermore, the result from a study by Szymanski et al indicate it was quantity of urinary leakage that was the most bothersome (92). Perrouin-Verbe discussed in their article the medical risk of operating on individuals with MMC because of their cognitive disturbances. Until adulthood, there is support from the parents in coping with CIC, regular bladder washout, but when the individuals have to manage independently major problems may occur. Before decision about continence surgery, the question is how to select those individuals with MMC who can cope independently after the operation. However, continence surgery is the only thing that provides complete urinary continence, which is important to a good life in the company of others. (90).

FUTURE PERSPECTIVES

Transition from pediatric to adult care

For adults with MMC, there is a great need for continuous follow-up (81,82,90,93). When transition from pediatric to adult care occurs, the process should be individually designed, and account must be taken of the individuals′ maturity and disability. NOBAB-standard (Nordic standard for children and young people in health care proclaims that, at the time of the transition to adult care, the adolescent has the right to continuity, preparation, information, cooperation, respect, integrity and qualified personnel (94). Adult health care is not organized to meet these multi-medical needs (95-97). TRAQ-SB (Transition Readiness Assessment Questionnaire Spina Bifida) is used in many countries to evaluate whether the individual is mature enough to be transferred to adult care (98). Roth et al found out that only older age was associated with transition readiness (99). A successful organization according to transition readiness for adolescents with MMC could be a closer cooperation between pediatric and adult care. However, there is still questions raised about how to overcome the difficulties for this patient group described in examples given by the researchers; The adult ward was just around the corner from pediatric ward but only 40% attended the adult care (100). Roth et al asked; do we demand too much of the young people with MMC? (101). Hettel et al found out that even with new urological problems the adults did not seek care in the adult ward (102).

In the dissertation, we have not focused on fecal incontinence, but we are fully aware that it is a major problem for individuals with MMC. In tables in Studies I and III, we have shown the frequency of fecal incontinence. In Study IV, questions about experience of fecal incontinence were also asked, but the results will be presented in a further study. Szymanski et al evaluated the impact of fecal incontinence on HRQoL in children from the age of 8, as well as in adolescents and adults. Results showed that the quality of life was impaired (103).

6 CONCLUSION

In adults with MMC from western Sweden, born between 1957 and 1980, about 60% were single and just a few had children. It seemed to be easier for females to establish close relations. Few had urotherapy support, none had support with their fecal elimination regimen and the majority used pads. To use pads had a negative influence on becoming employed.

To evaluate HRQoL. SF-36 was used. As an overall group they had significant impairment of PCS and improvement of MCS compared to the Swedish population. Surprisingly, HRQoL was unaffected by fecal incontinence, partnership and offspring. Even in the individuals who were dysphoric during the interview, MCS was unaffected, while PCS was decreased.

As we believe SF-36 to be an unreliable instrument, a personal interview with a qualitative approach was performed in 9 of the individuals who had urinary incontinence and who could openly recount how incontinence restricts and affects their lives. The results are consistent with reports from individuals with incontinence due to acquired conditions.

With standardized follow-up, active treatment strategy, and urinary/bowel therapy, almost 70% of adolescents with MMC can reach urinary continence. Urinary continence seems to be a success factor in creating an age-appropriate active social life and seems to enhance the possibility of finding a partner and becoming physically intimate.

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APPENDIX

1.The questionnaire for study I in Swedish.

2.The questionnaire for study III in Swedish.

1.The questionnaire for study I in Swedish.

Grundläggande data	Namn Personnummer Tidigare aktuell barnhabilitering
Skadenivå enligt barnjournalen	1 = cervikalt 2 = thorakalt 3 = thorakolumbalt 4 = lumbosakralt 6 = sakralt
Hydrocefalus enligt barnjournalen	0 = ingen hydrocefalus 1 = opererad hydrocefalus 2 = hydrocefalus ej opererad (om data finns)
Intervjufrågor	
Förflyttningssätt	0 = utan hjälpmedel men kan ha ortopediska inlägg/skor 1 = skenor/kryckor (vanligen) 2 = delvis rullstol 3 = alltid rullstol
Skolgång/utbildning	Högsta utbildning 1 = särskola inklusive särgymnasium 2 = normalklass grundskola 3 = gymnasium 4 = folkhögskola 5 = universitet
Nuvarande sysselsättning	0 = sysslolös 1 = dagcenter/annan liknade lösning 2 = studier 3 = lönearbete
Genomgått körkortsutbildning	0 = nej 1 = ja
Boendeform	1 = eget boende 2 = eget boende i servicehus 3 = gruppboende 4 = föräldrahem
Assistent	0 = nej 1 = ja om ja, vad gör assistenten?

Civilstånd	1 = single 2 = sambo/gift
Sexuella erfarenheter	0 = inga 1 = ja om ja, ”vill du berätta om detta?”
Graviditeter	0 = inga 1 = ja; antal
Barn	0 = inga 1 = ja; antal
Fertilitetsförsök	0 = nej 1 = ja om ja, vilken typ av fertilitetsförsök
Urologiska frågor	
Blåstömningsmetod	1 = spontan/normal miktion 2 = krystning/komprimering 3 = RIK 4 = annan metod 0 = ingen (bara rinner)
Självständighet i samband med blåstömning	0 = nej ”vad behöver du hjälp med?” 1 = ja (klarar allt)
Antal tömningar/dygn	0 = ingen (bara rinner) 1 = 1-3 2 = 4-6 3 = >6
Urinläckage, dag	0 = inget läckage 1 = enstaka läckage/månad 2 = enstaka läckage/vecka 3 = enstaka läckage/dag 4 = alltid våt
Förbrukningsartiklar	
Katetertyp	1 = obehandlad 2 = behandlad 3 = förgelad 4 = annan

Kateterstorlek, Ch	1 = Ch8 2 = Ch10 3 = Ch12 4 = Ch14 5 = Ch16 6 = annan, vilken
Kontinensskydd	0 = nej 1 = ja om ja, vilken typ
Toaletthjälpmedel	0 = inget 1 = ja om ja, vilken typ
Tarmtömningsmetod	1 = spontan/normal tarmtömning 2 = krystning 3 = tarmsköljning om ja Resulax Klysma Vatten Blandning vilken 4 = annan metod vilken
Avföringsläckage, dag	0 = inget läckage 1 = enstaka läckage/månad 2 = enstaka läckage/vecka 3 = enstaka läckage/dag 4 = alltid avföringsläckage
Avföringsläckage, natt	0 = inget läckage 1 = enstaka läckage/månad 2 = enstaka läckage/vecka 3 = enstaka läckage/dag 4 = alltid avföringsläckage
Självständighet i samband med tarmtömning	0 = nej om nej, ”vad behöver du hjälp med?” 1 = ja
Frekvens	0 = ingen 1 = 1 ggr/vecka 2 = 2 ggr/vecka 3 = varannan dag 4 = varje dag 5 = annat

Förbrukningsartiklar av avföringsinkontinens	0 = inga 1 = lavemangspip/påse 2 = tarmtratt/påse eller behållare 3 = ballongkateter/påse 4 = annat
Särskilt inkontinensskydd	0 = nej 1 = ja om ja, vilken typ
Aktuella läkemedel	0 = inga 1 = urologiska läkemedel vilka 2 = övriga läkemedel vilka
Genomgången kirurgi	0 = ingen 1 = ja blåsa vilken tarm vilken
Behandlande UVI:er	0 = nej 1 = ja ofta >1 ggr/år färre < 1 ggr/år
Senaste urologiska kontroll	0 = ej sedan överföringen 1 = årtal
Uroterapeutkontakt	0 = nej 1 = ja var

2.The questionnaire for study III in Swedish.

Grundläggande data	Namn Personnummer Barnhabilitering
Skadenivå enligt journal	Cervikalt Thorakalt Thorakolumbalt Lumbalt Lumbosakralt Sakralt
Hydrocefalus	Ingen Op hydrocefalus Ej op hydrocefalus
Förflyttning	Utan hjälpmedel Kryckor Delvis rullstol Alltid rullstol
Utbildning	Särskola Grundskola Särgymnasium Gymnasium
Körkort	Ja Nej
Boende	Eget Eget i servicehus Gruppboende Föräldrahem
Assistans	Ja Nej om ja, vad gör assistenten?
Urologiska frågor	
Blåstömningsmetod	Spontan Krystning RIK Annan metod Ingen

Antal tömningar/dygn	0 1-3 4-6 >6
Självständig	Ja Nej om nej, vad behöver du ha hjälp med?
Vid RIK – katetertyp	Obehandlad Hydrofil Förgelad Annan
Kateterstorlek/Ch	Ch8 CH10 Ch12 Ch14 Ch16
Annan typ eller storlek	Ch18 Annan sort, vilken?
Urinläckage, dag	Inget Enstaka/månad Enstaka/vecka Enstaka/dag Alltid våt
Urinläckage, natt	Inget Enstaka/månad Enstaka/vecka Enstaka/dag Alltid våt
Kontinensskydd	Ja Nej om ja, vilken sort?
Toaletthjälpmedel	Ja Nej Vad?

Tarmtömning	
Metod	Spontant Krystning Tarmsköljning med vad? Annan metod Hur ofta?
Hjälpmedel vid tarmtömning	Vilket?
Avföringsläckage/dag	Inget Enstaka/månad Enstaka/vecka Enstaka/dag Alltid
Avföringsläckage/natt	Inget Enstaka/månad Enstaka/vecka Enstaka/dag Alltid
Självständighet	Ja Nej om nej, vad behöver du hjälp med?
Kontinensskydd enbart för avföringsläckage	Ja Nej om ja, vad för sort?
Övriga frågor	
Aktuella läkemedel/sort	Inga Urologiska Övriga
Genomgången kirurgi	Ja Nej Blåsa Tarm
Behandlade UVI:er	Ja Nej Hur ofta?
Senaste urologiska kontroll	Årtal

Kontakt med uroterapeut	Ja Nej
Genomgått psykologtest	Ja Nej
Kognitiv nivå enligt utlåtandet	
Nära intima relationer/erfarenheter? Sexuella erfarenheter?	Ja Nej om ja, vill du berätta?
Fritidsaktiviteter/kamratkontakt? Beskriv kamratkontakten. Hur många? Vilken typ av relation?	Ja Nej
Lätt att ta/få kontakt med andra? Vem umgås du helst med? Sover du över hos någon? Idrottar du? Har du något annat fritidsintresse?	Ja Nej
Om kontinensskirurgi, hur var det före/efter op angående idrott/andra fritidsintressen/umgås med andra/övernatta?	Ja