Acta Pædiatrica ISSN 0803–5253

REGULAR ARTICLE

Medical problems in adolescents with myelomeningocele (MMC): an inventory of the Swedish MMC population born during 1986–1989

I Olsson (ingrid.b.olsson@vgregion.se)[1], M Dahl[2], S Mattsson[3], M Wendelius[4], E Åström[5], L Westbom[6]

1.Department of Paediatrics, Sahlgrenska University Hospital, Göteborg, Sweden
2.Department of Women's and Children's Health, University Hospital, Uppsala, Sweden
3.Department of Paediatrics, University Hospital, Linköping, Sweden
4.Department of Paediatrics, Norrland University Hospital, Umeå, Sweden
5.Department of Paediatrics, Karolinska University Hospital/Solna, Stockholm, Sweden
6.Department of Paediatrics, University Hospital, Lund, Sweden

Keywords
Adolescence, Adult, Comprehensive care, Myelomeningocele, Spina bifida

Correspondence
I Olsson, Department of Paediatrics, The Queen Silvia Children´s Hospital,
S-416 85 Göteborg, Sweden.
Tel: +46 31 343 40 00 | Fax: +46 31 25 79 60 |
Email: ingrid.b.olsson@vgregion.se

Received
27 April 2006; revised 15 September 2006; accepted 16 November 2006.

DOI:10.1111/j.1651-2227.2006.00153.x

Abstract

Aim: To describe the prevalence of myelomeningocele (MMC) and the medical needs of adolescents, 15–18 years, with MMC in Sweden, at a time when they are on the threshold of adulthood, leaving paediatrics.

Methods: In a retrospective study, we identified all adolescents with MMC, born during 1986–1989 and living in Sweden on July 1, 2004. An inventory was agreed upon with questions concerning their medical problems and need for medical care.

Results: There were 175 persons 15–18 years of age, born with MMC or lipoMMC (prevalence 3.8 per 10 000). Hydrocephalus was seen in 86%, 31% had been operated because of tethered cord syndrome, and 6% for Chiari malformation symptoms. The majority had motor impairments. Clean intermittent catheterisation for bladder emptying was used by 85%, and 59% used enemas on a regular basis because of the neurogenic bowel dysfunction. Renal dysfunction was seen in 1.7% of the adolescents.

Conclusion: Lifelong follow-up by many specialists, among others neurologists and neurosurgeons, urotherapists and urologists, orthopaedic surgeons and orthotists, is necessary for individuals with MMC. The complex medical situation, often in combination with cognitive difficulties, makes it necessary to coordinate medical services for this increasing group of adults with multiple impairments.

INTRODUCTION

The prognosis for children born with myelomeningocele (MMC) is much better today than it was 40 years ago when only 40% reached the age of 7 years (1). Even in the early 1970s, the mortality rate before the fifth birthday was 34% (2). Cardiorespiratory failure, most likely due to Chiari malformation, was one cause of death in young age. Hydrocephalus, shunt problems and central nervous system (CNS) infections were other causes of death (2,3). Better shunting procedures and brainstem decompression have contributed to the decreasing mortality rates in early childhood (4,5). Renal failure and shunt complications are still the most important causes of death. Due to better treatment of hydrocephalus and the neurogenic bladder dysfunction, the survival rates in early adulthood have increased from less than 50% (2,3) to about 75% (6). The medical problems, however, persist throughout life and involve the urinary tract (infections, stones, renal failure), the CNS (shunt complications, infections, tethered cord syndrome, Chiari-related symptoms) and the skin (pressure sores) (2,6–8). Latex allergy, common in individuals with MMC, may lead to life-threatening events (9).

The motor impairment may become more obvious due to various factors, such as poor physical condition and overweight, and many who walked ≥50 metres as children cease to do so as adults (2). Neurological deterioration due to the tethered cord syndrome and Chiari malformation may also worsen the motor impairment (6,10). Shunt dysfunction is a constant threat and can cause acquired brain injury and blindness in addition to the existing impairments (2). Endocrinological dysfunction such as precocious puberty is common (2). In addition, there are reports on early ageing in individuals with MMC (11). The cognitive impairments in persons with MMC and hydrocephalus are well documented (12–17). Many have difficulties in taking initiative, planning and organising activities, which is a major handicap at the labour market and in society (12,14,16). This impairment of executive function also explains the urgent need for coordinated health care for adults with MMC (18).

In Sweden, 20–25 children are born with MMC every year (19). Initially, they are taken care of at one of the six regional (tertiary) centres with neuropaediatric and neurosurgical units (at one centre paediatric surgeons by tradition do the initial operations). The children have their medical check-ups and habilitation at the local level, where neuropaediatricians coordinate the various medical specialist interventions. The regional units carry out regular controls of bladder and renal function throughout childhood and adolescence. The majority reach adulthood, including those

with severe disabilities. Despite the fact that many have severe medical problems, there is a lack of coordinated medical care for adults with MMC in Sweden.

The aim of the present study is to investigate the medical needs in the group of adolescents about to reach adulthood. It is made as a part of a new collaboration between the neuropaediatricians at the regional centres of Sweden, which aims at setting guidelines for the medical care in MMC.

METHOD

All adolescents with MMC born during 1986–1989 and living in Sweden on July 1, 2004 were identified, including those with lipomyelomeningocele (lipoMMC) whose medical problems are similar to those of open MMC. Each of the authors scrutinized the medical files at the regional centres, and when needed the local habilitation units were contacted. The retrospective nature of the study made us agree on a minimum of questions to cover the most important issues and where information could be achieved about all the adolescents (Table 1).

RESULTS

On July 1, 2004, 175 adolescents with MMC born between 1986 and 1989 lived in Sweden. The group consisted of 89 boys and 86 girls. Nine of them had lipoMMC. The prevalence of MMC including lipoMMC at ages 15–18 years was 3.8/10 000.

The majority had hydrocephalus (86%), 2/9 with lipoMMC and 148/166 (89%) with open MMC. Seven had been operated with endoscopic ventriculocisternostomy, two of whom needed also a ventriculoperitoneal shunt. In all, three adolescents had serious deficits (visual and motor) after shunt complications. Eleven (6%) had Chiari-related symptoms leading to a brainstem decompression. Three of five who needed mechanical ventilation received treatment 24 h a day. Six had a gastrostomy due to feeding difficulties. Learning disabilities were common with 27% having an IQ level <70. Five per cent had a severe disability with an IQ level <50. Fourteen per cent had active epilepsy.

Most had a severe motor impairment: 53% did not walk at all indoors, and another 20% used walking aids indoors.

Table 1 Issues investigated

Hydrocephalus	y/n	Neurosurgical procedure	
Chiari-related symptoms	y/n	Decompression	y/n
Need for mechanical ventilation	y/n	Gastrostomy	y/n
Motor impairment		Walking indoors	y/n
Use of orthoses	y/n	Spontaneous fractures	y/n
Scoliosis, treated	y/n	Scoliosis, operated	y/n
Tethered cord release	y/n	Frequent pressure sores	y/n
Cognitive level IQ<70, IQ< 50	y/n	Precocious puberty	y/n
CIC	y/n	Medication	y/n
Renal dysfunction	y/n	Urological operation	y/n
Mode of bowel emptying		Retrograde or antegrade enema	
Use of incontinence pads	y/n		

y/n: yes/no; CIC: clean intermittent catheterisation.

About 75% used orthoses. Spontaneous fractures had occurred in 11%, and 40% had needed treatment for scoliosis (operation or brace). The majority (75%) needed regular follow-up by an orthopaedic surgeon and orthotists.

The number of adolescents who had undergone tethered cord release varied between the regions, 31% in the whole population (15–65%). There were also regional differences in the number of operations for scoliosis (9–40%, 25% in the whole population), with the lowest proportion of operations in the region with the highest proportion of untethering procedures.

Sensory deficits, in combination with impaired mobility, led to increased risk of pressure sores, and 19% of the adolescents had frequent pressure sores.

Endocrinological disturbances were overrepresented in the group and in 10% the puberty was precocious. Weight problems were common and even if overweight dominated, the opposite was also found.

The majority (85%) used clean intermittent catheterisation (CIC) for bladder emptying. Pharmacological treatments of bladder dysfunction were used by 40%. Fifteen per cent had recurring urinary tract infections, and 21% used prophylactic antibiotic treatment. Only 3/175 (1.7%) had decreased renal function. Bladder enlargement or urinary deviations had been performed in 24% of the adolescents. Even if many of the adolescents achieved bladder control, they used incontinence pads for safety reasons–69% used them at least occasionally.

The neurogenic bowel dysfunction is usually treated with enemas on a regular basis; retrograde colonic washouts were used by 59%. Ten per cent used Malone antegrade colonic enemas (MACE). Thirty-five per cent needed incontinence pads due to incomplete bowel control. In all, 71% needed some kind of incontinence pads.

Only two adolescents (1%) were considered to manage without regular medical check-ups in adulthood due to their MMC.

DISCUSSION

All children with MMC born in Sweden are treated at one of the six regional centres in the neonatal period for closure of the cele and for shunting operation, and usually also for the specialised medical care in childhood and adolescence, and they are regularly followed by neuropaediatricians at the local child habilitation units. Thus, we believe we have found all 15- to 18-year-old adolescents with MMC living in Sweden on July 1, 2004, and they are likely to be representative of individuals with MMC born in the late 1980s. The drawback of this study is its retrospective nature, which implies that detailed information could not be obtained, but we still think it serves its purpose: to point at the medical needs of young adults with MMC.

The prevalence of MMC in this age group was 3.8/10 000. According to the Swedish register of malformations, the incidence of MMC during 1986–1989 varied between 3.5 and 5/10 000 (19). We do not know the birth and childhood mortality rate for our study group.

According to international reports, the dominant cause of death has been renal failure in individuals with MMC up to now (2,8). Renal impairment has been reported to occur in as many as nearly half of the adults with MMC (20) and is a common cause of hospitalisation in adults (6,7), as are urinary tract infections. The most striking finding in our series was that only three adolescents had renal dysfunction, probably due to the good urological and urotherapeutical follow-up in our country, and the national guidelines for neurogenic bladder dysfunction (21). In order to prevent infections and renal damage, but also in order to optimise continence (22,23), the regular evaluations of CIC technique and routines, of bladder function and of renal function must continue throughout life and not come to an abrupt end when the children become adults. Specialist care by urologists and urotherapists well acquainted with these problems is important (23,24). The aim to achieve complete bladder and bowel control can only be achieved in about 40% and 65%, respectively (23). We do not know the corresponding figures in our series, but the need for incontinence pads is obvious in about two-thirds, more as a measure of safety for many of the adolescents. Regular bowel enemas are important to obtain optimal continence, but not all are able to perform this without aid (25).

The percentage of individuals needing cerebral spinal fluid diversion is in accordance with other reports (6). We have not counted the number of shunt revisions in this population, but many needed several revisions, and in the study by Bowman et al., 95% needed at least one revision (6). Signs and symptoms of shunt dysfunction and shunt infection must be recognized early, since a delay may lead to additional impairments or even have lethal consequences (2). There is a risk of progressing neurological symptoms due to the tethered cord syndrome, the Chiari malformation and the development of hydromyelia. Continuous neurological assessments are therefore needed (6,7,10).

The need for orthopaedic check-ups and continuous orthopaedic technical support is obvious. Treatment of pressure sores in an individual with sensory impairments requires specialist care of a nurse acquainted with the problem, otherwise there is a risk of deeper infection or septicaemia or even amputation (18).

We cannot explain the regional differences when it came to tethered cord release and operations for scoliosis. Other orthopaedic surgical procedures were performed at all levels of health care making it impossible to gather all the necessary data in this retrospective study. Our impression was that the orthopaedic treatment tradition varied between the regions, which calls for more evidence-based approach to orthopaedic and neurosurgical interventions.

The rate of learning disabilities was high in this population of adolescents with MMC, but we cannot report details on their cognitive level. We know from other studies of cognitive function of individuals with MMC and hydrocephalus that there is an overrepresentation in the IQ range 70–80 (17), which also implies difficulties in mainstream schools and during daily activity. Adults with MMC have been shown to have poor numeracy skills (13) and deficient reading comprehension and writing (15), which also negatively effects independence in adult life. There seems to be a correlation with the number of shunt revisions, which points at the importance of minimizing the number of episodes with increased intracranial pressure (26).

There is an increased risk of latex allergy in individuals with MMC, which professionals must be aware of, and all operations on individuals with MMC should be latex-free. We lack data about the size of the problem in our population, but it has been reported in every third to fifth person with MMC (2,6,9). We also have insufficient data about endocrinological and weight problems in our population, which shows the need for more thorough assessment during adolescence.

The aim of this report was to focus on the complexity of medical problems in young adults with MMC, which calls for comprehensive care throughout life and close collaboration between several specialists. At the transition from childhood to adulthood, the young individual with MMC needs to know where to turn to within the health care system. The practical problems for the person with MMC might otherwise be overwhelming, leading to considerable medical risks (18). A malfunctioning shunt may add to the cognitive difficulties already existing, thereby preventing the individual to realize the imminent need for neurosurgical intervention.

In their 25-year follow-up of children with MMC, Bowman et al. conclude that it is one of the greatest challenges in medicine today to establish a network of care for adults with MMC (6). We fully agree.

ACKNOWLEDGEMENTS

The authors are grateful to Dr Bo Ericsson and Dr Ingegerd Åhsgren for contributing data to the study and to Dr Kate Abrahamsson for urological expertise. We are also most thankful to the dedicated urotherapists at the regional centres for their contribution to the data collection, and to the staff at the local habilitation centres in Sweden.

References

1. Lorber J. Results of treatment of myelomeningocele. An analysis of 524 unselected cases with special reference to possible selection for treatment. *Dev Med Child Neurol* 1971; 13: 279–303.
2. Hunt GM, Oakeshott P. Lifestyle in adults aged 35 years who were born with open spina bifida: prospective cohort study. *Cerebrospinal Fluid Res* 2004; 1: 4. Available from: http://www.cerebrospinalfluidresearch.com/content/1/1/4
3. Hunt GM, Oakeshott P. Outcome in people with open spina bifida at age 35: prospective community based cohort study. *BMJ* 2003; 326: 1365–6.
4. Dahl M, Ahlsten G, Carlson H, Ronne-Engström E, Lagerkvist B, Magnusson G, et al. Neurological dysfunction above cele level in children with spina bifida cystica. A prospective longitudinal study up to three years of age. *Dev Med Child Neurol* 1995; 37: 30–40.
5. Worley G, Schuster JM, Oakes WJ. Survival at 5 years of a cohort of newborn infants with myelomeningocele. *Dev Med Child Neurol* 1996; 38: 816–22.
6. Bowman RM, McLone DG, Grant JA, Tomita T, Ito JA. Spina bifida outcome: a 25-year prospective. *Pediatr Neurosurg* 2001; 34: 114–20.

7. Kinsman SL, Doehring MC. The cost of preventable conditions in adults with spina bifida. *Eur J Pediatr Surg* 1996; 6(Suppl 1): 17–20.
8. Müller T, Arbeiter K, Aufricht C. Renal function in myelomeningocele: risk factors, chronic renal failure, renal replacement therapy and transplantation. *Curr Opin Urol* 2002; 12: 479–84.
9. Rendeli C, Nucera E, Ausili E, Tabacco F, Roncallo C, Pollastrini E, et al. Latex sensitation and allergy in children with myelomeningocele. *Child Nerv Syst* 2006; 22: 28–32.
10. Craig JJ, Gray WJ, McCann JP. The Chiari/hydrosyringomyelia complex presenting in adults with myelomeningocele: an indication for early intervention. *Spinal Cord* 1999; 37: 275–8.
11. Dillon CM, Davis BE, Duguay S, Seidel KD, Shurtleff DB. Longevity of patients born with myelomeningocele. *Eur J Pediatr Surg* 2000; 10(Suppl 1): 33–4.
12. West M, Fjeldvik L, Rand-Henriksen S. Helping to solve problems associated with spina bifida. *Eur J Pediatr Surg* 1995; 5(Suppl 1): 12–5.
13. Dennis M, Barnes M. Math and numeracy in young adults with spina bifida and hydrocephalus. *Dev Neuropsychol* 2002; 21: 141–55.
14. Barf HA, Voerhoef M, Jennekens-Schinkel A, Post MW, Gooskens RH, Prevo AJ. Cognitive status of young adults with spina bifida. *Dev Med Child Neurol* 2003; 45: 813–20.
15. Barnes M, Dennis M, Hetherington R. Reading and writing skills in young adults with spina bifida and hydrocephalus. *J Int Neuropsychol Soc* 2004; 10: 655–63.
16. Iddon JL, Morgan DJ, Loveday C, Sahakian BJ, Pickard JD. Neuropsychological profile of young adults with spina bifida with or without hydrocephalus. *J Neurol Neurosurg Psychiatry* 2004; 75: 1112–8.
17. Lindquist B, Carlsson G, Persson E-K, Uvebrant P. Learning disabilities in a population-based group of children with hydrocephalus. *Acta Paediatr* 2005; 94: 878–83.
18. Kaufman BA, Terbrock A, Winters N, Ito J, Kosterman A, Park TS. Disbanding a multidisciplinary clinic: effects on the health care of myelomeningocele patients. *Pediatr Neurosurg* 1994; 21: 36–44.
19. National Board of Health and Welfare. Medical birth registration. Available from: http://www.sos.se/epc/fodelse/mfrfiler/nationell.htm
20. McDonnell GV, McCann JP. Issues of medical management in adults with spina bifida. *Child's Nerv Syst* 2000; 16: 222–7.
21. Gladh G, Mattsson S. (red) Nationellt vårdprogram för barn med neurogen blås- och tarmstörning (*The Swedish national programme for children with neuropathic bladder and bowel*). Stockholm: Socialstyrelsen (National Board of Health and Welfare), ISSN 1403-3348, 1998.
22. Lapides J, Diokno AC, Silber SJ, Lowe BS. Clean, intermittent self-catheterization in the treatment of urinary tract disease. *J Urol* 1972; 107: 458–61.
23. Verhoef M, Lurvink M, Barf HA, Post MW, van Asbeck FW, Gooskens RH, et al. High prevalence of incontinence among young adults with spina bifida: description, prediction and problem perception. *Spinal Cord* 2005; 43: 331–40.
24. Borzyskowski M. An update on the investigation of the child with a neuropathic bladder. *Dev Med Child Neurol* 1996; 38: 744–8.
25. Mattsson S, Gladh G. (2005) Tap water enema for children with myelomeningocele and neurogenic bowel dysfunction. *Acta Paediatr* 2006; 95: 369–74.
26. Hetherington R, Dennis M, Barnes M, Drake J, Gentili F. Functional outcome in young adults with spina bifida and hydrocephalus. *Child's Nerv Syst* 2006; 22: 117–24.