

The educational journey of individuals with MPS II Hunter Disease in the United Kingdom^(B)

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Introduction

- Hunter disease (MPS II) is the only known X-linked MPS disorder.¹
- The deficiency of the lysosomal enzyme iduronate-2-sulphatase leads to a progressive accumulation of glycosaminoglycans in the body and an array of clinical manifestations, including skeletal and cardiac abnormalities.¹
- Traditionally, individuals were classified as 'mild' or 'severe', based on the absence or presence of central nervous system (CNS) involvement; it is now recognised that the syndrome exists in a range somewhere between the two extremes, now classified as attenuated to severe.^{1,2}
- Around two-thirds of individuals are estimated to have the severe phenotype (i.e. progressive CNS involvement); these individuals experience learning difficulties and neurological decline.^{1,2}
- The aim of this project was to determine the cognitive variability in patients with MPS II in the United Kingdom (UK) and to understand their needs and support requirements in an educational setting.

Methods

- Seventy-one individuals with MPS II resident in the UK were identified by the MPS Society and invited to take part in the survey via telephone interview.
- A specifically designed questionnaire was used to assess the individuals' diagnoses, treatment, educational attainment and need for support from primary through to further education.
- Interviews took place in December 2015 and January 2016.
- Results for the education section of the survey only are presented here.
- Results for the diagnosis and treatment section of the survey are presented in Poster 165.

Results

- Forty-one individuals agreed to take part in the study (58%), ranging in age from 1 to 36 years (mean 12.3 years).
- Of the 41 individuals surveyed, 54% (n=22) reported CNS involvement; 37% (n=15) reported no CNS involvement; and 4 individuals (10%) did not know whether there was CNS involvement or not.
- A review of all respondents data indicated all but 3 had some level of CNS involvement; 49% (n=20) had severe progressive CNS involvement.
- One patient, aged 1 at questionnaire completion, was too young to have attended nursery/primary school and is excluded from this analysis.
- Most individuals started their education in a mainstream school (Figure 1); one third of individuals (n=13) moved primary schools as their learning needs were not being met; individuals moved school at a mean age of 6.8 years.
- Three quarters of the individuals who moved primary schools were originally in a mainstream school (n=10) (Figure 1); of these, 8 moved to a special educational needs school (SEN); 1 individual moved to a mixed school, 1 did not answer.
- All individuals who moved from a primary SEN school (n=3) moved to an alternative SEN school.
- Individuals with central nervous system (CNS) involvement were more likely to move school than those without (55% vs 7%).
- Nineteen individuals had attended or were attending secondary school (Figure 1); 1 individual moved from a secondary SEN school to mainstream secondary school at age 12.
- Statements of educational need or educational healthcare plans (EHPs) were issued to 73% of individuals in primary school (mean age 4.9 years).
- The reasons for issuing statements or EHPs in primary school were learning needs (15%), physical needs (5%) or both (52%).
- Fewer individuals (68%) had statements or EHPs during their secondary education; 23% of these had been issued at secondary school (mean age 11.3 years).
- The reasons for issuing statements or EHPs in secondary school were learning needs (7%), physical needs (15%) or both (53%).
- More individual education plans (73% vs 42%) and flexible teaching (57% vs 40%) were available in secondary compared to primary schools.
- Flexibility included alternatives to physical education (PE) lessons (26%), support lessons, and options to drop a GCSE in mainstream schools; and totally individual lesson plans in SEN schools.
- A breakdown of the support provided in primary and secondary schools is shown in Figures 2 and 3, respectively.

Figure 1. Primary and secondary education.

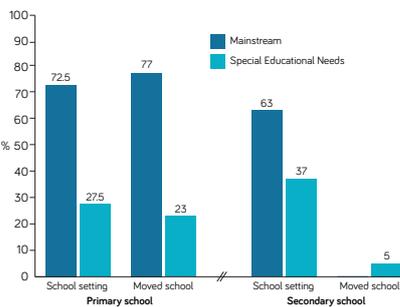


Figure 2. Support provided in primary school

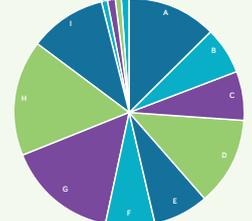
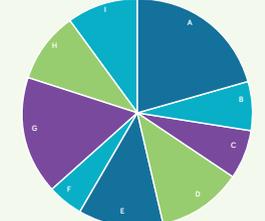


Figure 3. Support provided in secondary school



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Figure 7. Employment history of individuals with MPS II

*Some individuals reported more than one role

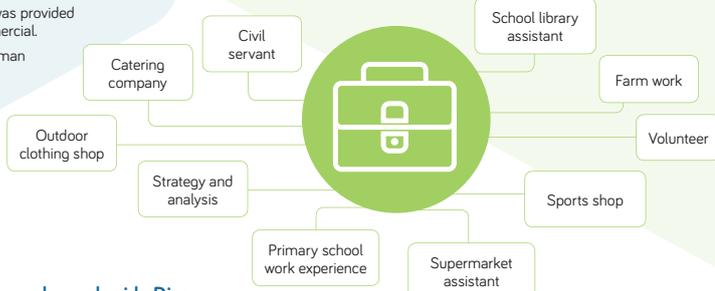


Figure 4. Professional input in primary school

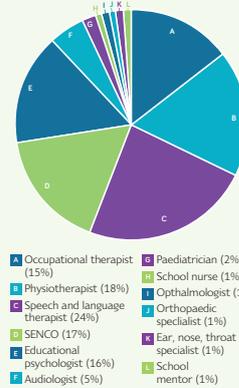
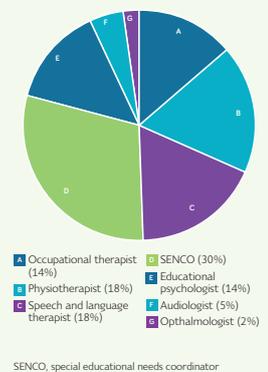
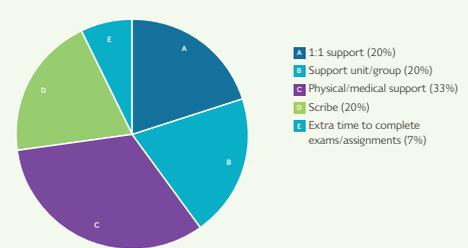


Figure 5. Professional input in secondary school



- Sixty three percent of individuals felt that their support needs had changed from primary to secondary education; with reasons cited as a decline in mobility/more help to move around larger schools (31%), difficulty understanding work (57%), and deteriorating health/surgery (15%).
- Of the 13 individuals aged 16 or over, 69% had obtained GCSE or equivalent qualifications.
- Ten individuals were attending or had attended further education including 6th form, college and university; all received some degree of support throughout their further education, a breakdown of which is shown in Figure 6.

Figure 6. Support received in further education



- Individuals achieved a range of qualifications from 'A' levels through to Master's degree.
- Of the 8 individuals who had completed their education 50% were working in voluntary or paid employment in a variety of roles* (Figure 7).

Conclusions

- Our findings report a lower prevalence of 'severe' phenotype than published estimates.²
- In the UK, there is a wide range of support available for individuals with MPS II in both SEN and mainstream education.
- Educational needs for those with CNS involvement cannot always be met in mainstream primary schools, but this is usually recognised, and more suitable SEN schooling found by age seven.
- Support requirements and the need for a flexible approach appear to change from primary to secondary school due to disease progression, the demands of moving around a larger school and the difficulty of the school work.
- Half of the MPS II sufferers surveyed who had completed their education had gained further education qualifications and found employment in the voluntary or paid sectors.

References

- Wraith J, et al. 2008. Eur J Pediatr;167:267.
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