



MULTIDISCIPLINARY MANAGEMENT OF CHILDREN WITH PRADER-WILLI SYNDROME IN THE FRENCH DATABASE OF PWS

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INTRODUCTION: The reference centre for Prader-Willi syndrome, set-up by the health ministry in November 2004, involves 6 regions of the South of France with a coordination team in Toulouse. One of the objectives of the centre is to set-up a database starting with children and adolescents containing medical and psychosocial data. This database is partially supported by Pfizer Inc.

METHODS: One hundred and forty eight children (81 male, 67 female) were included in the database with a median chronological age at inclusion of 7.3 years [0.21-23.25]. Median age at diagnosis was 3 months [0.1-144] with 64% deletion and 24% uniparental disomy. The objective of the study was to evaluate the multidisciplinary management of these patients (nutritional management, physiotherapy, psychomotricity, speech and language management) at different ages (younger than 6 years, between 6 and 12 years and older than 12 years). Data were collected at the entry in the database and yearly. We only present baseline data. Case report forms were filled by the paediatric endocrinologists and sent to our centre, which enters the data. The questions asked were if there was an active therapy or if the therapy was stopped or never proposed.

RESULTS:

	All	< 6 years	≥ 6 and <12 years	≥ 12 years
<i>Nutritional management by a dietician (N=137)</i>				
Yes, active	66.4%	78.6%	62.2%	52.8%
Yes, stopped	6.6%	1.8%	6.7%	13.9%
No, never proposed	27%	19.6%	31.1%	33.3%
<i>Physiotherapy (N=137)</i>				
Yes, active	52.5%	64.3%	44.4%	44.4%
Yes, stopped	38.7%	30.3%	44.4%	44.4%
No, never proposed	8.8%	5.4%	11.2%	11.2%
<i>Psychomotricity (N=135)</i>				
Yes, active	52.6%	65.5%	59%	25%
Yes, stopped	23%	3.6%	25%	50%
No, never proposed	24.4%	30.9%	16%	25%
<i>Speech and language management (N=139)</i>				
Yes, active	75.5%	82.1%	88.9%	50%
Yes, stopped	14.4%	0%	8.9%	42.1%
No, never proposed	10.1%	17.9%	2.2%	7.9%



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CONCLUSION: to our surprise, 27% of the patients did not have nutritional management by a dietician and 10% did not undergo speech and language therapy. More detailed analyses of these data are in progress. We could use this database to understand the difficulties of multidisciplinary management and to optimize it.