Overview of leading causes of death among French patients with Prader-Willi Syndrome, 2004-2014

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Category: Multisystem endocrine disorders

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Background
- Prader-Willi Syndrome (PWS) is one of the most common known causes of syndromic obesity, and is a major cause of morbimortality among this population.
- In the last 20 years, substantial improvements have been made regarding the diagnosis, treatment and management of patients with PWS.
- Few mortality data exist that take actual management into account and the creation of the French Reference Centre for PWS (FRC-PWS) in 2004 is a unique opportunity to investigate this issue.

Objective
- To report leading causes of mortality among the French patients with Prader-Willi Syndrome over eleven years of the nationwide FRC-PWS year mortality survey.

Methods
This study relied on two sources of mortality information at national level between 2004 and 2014:
- The CépiDc (French Epidemiological Centre for the Medical Causes of Death Registry):
  - Cases of death of patients with a PWS diagnosis (ICD code Q87.1: Congenital malformation syndromes predominantly associated with short stature).
- The FRC-PWS database:
  - Patients followed by endocrinologists from the 3 sites of the Reference Centre and the 22 Centres of Competence for PWS in France.
  - The information was corroborated with the French Prader-Willi Association.

Results
- Total number of cases: n=104
- 17 children
- 87 adults
- Number of cases of death per year

<table>
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<th>Year</th>
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<th>Adults</th>
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</tbody>
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- Median age at death for adults: 32.0 y/o (18.6;58.0)
- 70% of the children died within the first two years of life
- Respiratory related causes: more than 50% of deaths in patients with PWS

No significant differences were found by gender or genetic subtype regarding the causes or age of death.

Conclusion
- PWS is per se a condition that can result in premature death (median age at death for the total population: 30 y/o [0.1;58.0]).
- These findings highlight the respiratory vulnerability in PWS patients at all ages.
- Prevention and management of obesity and respiratory problems are the most important approaches to lower the mortality rate in this population.


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