

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

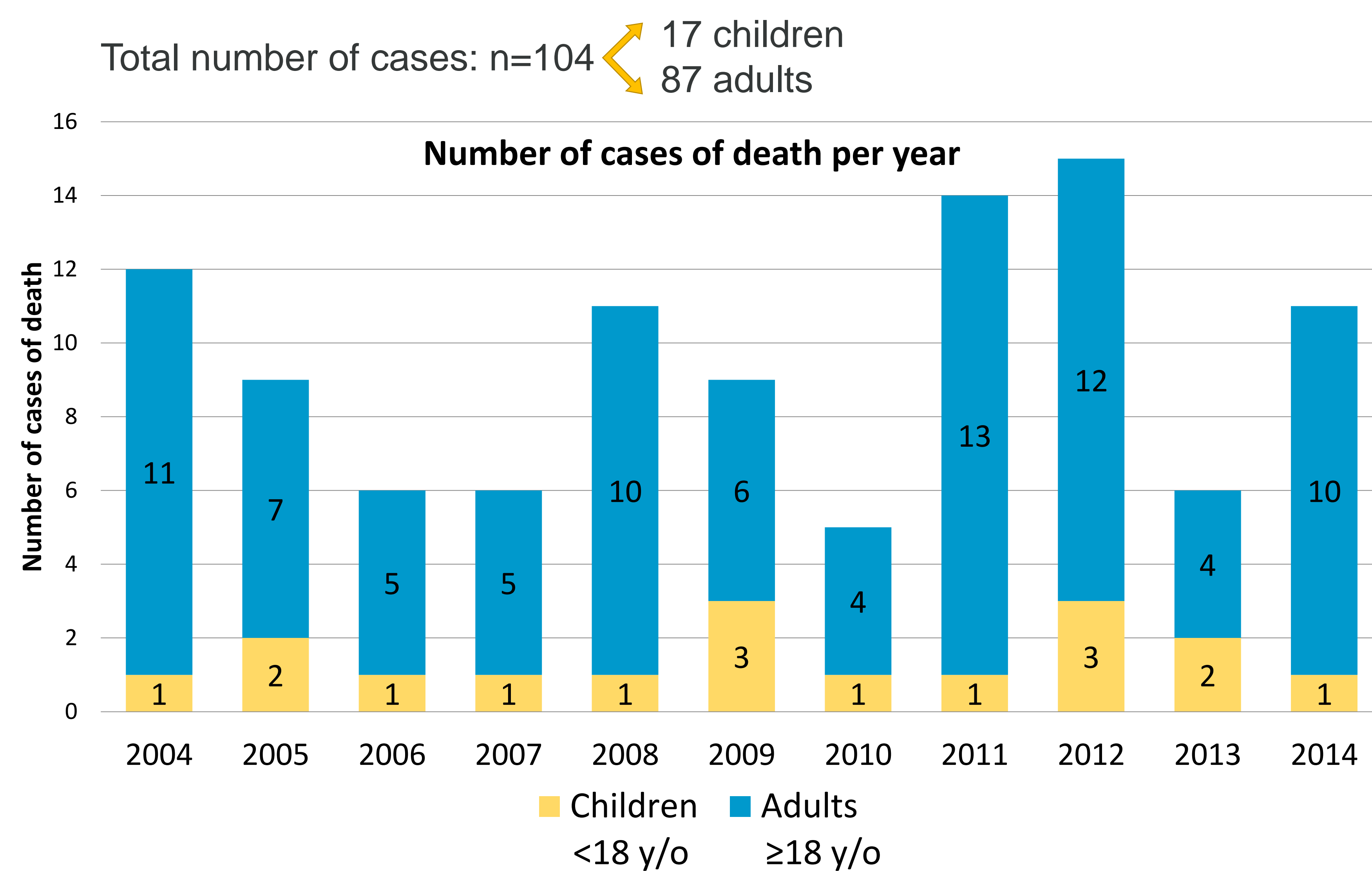
- To report leading **causes of mortality** among the French patients with Prader-Willi Syndrome over eleven years of the nationwide FRC-PWS

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



- **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

Primary causes of death :

- **Respiratory** ➤ n = 55
 - respiratory failure • 42 (40 adults)
 - respiratory infection • 13 (4 children)
- **Cardiovascular** ➤ n = 15
 - cardiac failure • 8
 - pulmonary embolism • 4
 - others cardiac cause • 3
- **Non-respiratory infection** ➤ n = 8
 - gastrointestinal infection • 2
 - sepsis • 4
 - others non-respiratory infection • 2
- **Sudden and unexplained death** ➤ n = 18
- **Unknown** ➤ n = 5
- **Others causes of death** ➤ n = 3

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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Multisystem endocrine disorders

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Poster presented at:

