

Defining an optimal time window to screen for hepatoblastoma in children with Beckwith-Wiedemann syndrome (2019)

Alessandro Mussa, Kelly A. Duffy, Diana Carli, Giovanni Battista Ferrero, Jennifer M. Kalish

Background

Patients with Beckwith-Wiedemann syndrome (BWS) are at an increased risk for embryonal tumors. Hepatoblastoma (HB), a rare liver tumor, represents the second most common tumor to develop in patients with BWS. To monitor for HB development, patients with BWS receive routine tumor surveillance until the age of 4 years and screening is continued until 7 years to monitor for other tumors.

Purpose

This study evaluated the ages at HB development in patients with BWS compared to the ages at HB development in patients without BWS to determine the optimal length of time to screen for HB in children with BWS.

Findings

A literature search was performed to identify all published reports of patients with BWS who developed HB and additional unpublished patients from the authors' registries were included. The ages at HB development in the patients with BWS was recorded and compared with a previously published group of patients with HB (referred to as the control cohort).

Patients with BWS often developed HB earlier than the control cohort:

	BWS Cohort	Control Cohort
Average age at diagnosis	7.1 ± 6.0 months	19.5 ± 14.7 months
Diagnosed before 12 months of age	80%	39%
Diagnosed before 18 months of age	95%	53%
Diagnosed before 24 months of age	97%	68%
Diagnosed before 30 months of age	100%	79%

Conclusion

All reported HB in patients with BWS developed before the age of 30 months. These data suggest that HB screening in patients with BWS may be reduced to until 30 months of age, although further research is needed before current recommendations should change. Current recommendations for HB screening in patients with BWS include serum alpha-fetoprotein (AFP) measurements and complete abdominal ultrasounds every 3 months until age 4 years. After age 4, HB screening can stop, however patients should receive renal ultrasounds every 3 months until age 7 years.

Key Points

- Patients with BWS develop hepatoblastoma earlier than the general population.
- HB in patients with BWS developed before 30 months of age.
- It may be possible to decrease the tumor screening age for HB, however further research is needed.

Reference

Mussa A et al. Defining an optimal time window to screen for hepatoblastoma in children with Beckwith-Wiedemann syndrome. *Pediatr Blood Cancer*. 2019;66(1): e27492. PubMed PMID: 30270492