

Hypothalamic hamartoma: epilepsy and neurodevelopmental profiles in a clinical cohort

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Clinical profiles

- Epilepsy and neurodevelopmental disorders, including autism, were highly prevalent.
- Epilepsy was associated with the presence of neurodevelopmental comorbidities.
- Autistic behaviours and neurodevelopmental concerns often arose first.
- Structural correlates for epilepsy and intellectual disability were identified.

Conclusions

- Seizures contribute to cognitive and behavioural dysfunction.
- The hamartoma itself is likely to be a primary determinant of the epilepsy clinical profiles.
- We postulate that the underlying aetiology, likely genetic, directly influences the clinical profile.
- Atypical neurodevelopmental profiles in HH could best be conceptualized as a developmental and epileptic encephalopathy.

Significance

- Structural characteristics, and presence or absence of epilepsy, can be used to predict the clinical course.
- This allows an individual approach to counselling, monitoring and consideration for early surgery.