

# Progressive myoclonic epilepsy

## □ Case Reports

In a single-patient illustrative case, **Mine et al.**, from Kyushu University, [Fukuoka](#), Japan, published in the [Journal of Neurosurgery Case Lessons](#), report the **first adult case** of [Dentatorubral-pallidoluysian atrophy](#) (DRPLA)-associated [progressive myoclonic epilepsy](#) (PME) treated with a **total corpus callosotomy (CC)** for [refractory seizures](#).

→ **Outcome:** Total CC led to a **marked reduction in seizure frequency**. Tonic seizures and FBTCSS with desaturation resolved by 1 year, with notable **improvement in quality of life (QOL)**.

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## □ Critical Appraisal

- □ **Strengths**

- Novelty: First reported adult DRPLA-PME corpus callosotomy.
- Clearly disabling epilepsy: Myoclonus, tonic seizures, desaturation.
- Clear outcome: Sustained seizure reduction at 1-year.

- □ **Limitations**

- Single case → low external validity.
- No comparator: No data vs. anterior CC or other therapies.
- Cognitive effects unquantified.
- DRPLA is a diffuse neurodegenerative disease; CC does not target focus directly.

- □ **Intellectual Rigour**

- Palliative intent is explicitly acknowledged.
- Cites prior PME cases (mostly pediatric, non-DRPLA).
- Lacks deeper comparison to alternatives (e.g. VNS, thalamic DBS).

## □ Clinical Verdict

★★★☆☆ **6/10** Interesting and rare case with clear illustrative value. However, it lacks broader context, comparative analysis, and rigorous outcome measurement.

## □ Takeaway for Practicing Neurosurgeons

**Clinical context:** In **adult patients** with [Dentatorubral-pallidoluysian atrophy](#) (DRPLA)-related **progressive myoclonic epilepsy (PME)** that is **disabling** and **refractory to standard therapies**,

→ a **total corpus callosotomy** may offer **clinically meaningful palliation**.

### Key benefits observed:

- **Rapid reduction** in seizure frequency
- **Resolution of desaturation** episodes
- **Improved quality of life (QOL)** at 1-year follow-up

### Safety:

- No major additional neurological or systemic harm reported postoperatively

**Bottom line:** > This case supports **considering total callosotomy** as a **palliative surgical option** in select adult PME cases where conventional therapies have failed.

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## Bottom Line

Total corpus callosotomy may be a **reasonable and effective palliative option** in **adult-onset DRPLA-related PME**, particularly when conventional treatments have failed.

## WordPress Categories

- Case Report
- Epilepsy Surgery
- DRPLA
- Corpus Callosotomy

## Tags

- PME
- DRPLA
- callosotomy
- adult epilepsy
- palliative neurosurgery
- case lessons

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Mine D, Shimogawa T, Sakai Y, Shigeto H, Okubo S, Sakata A, Watanabe E, Nakamizo A, Yoshimoto K. Total corpus callosotomy for an adult patient with progressive myoclonic epilepsy associated with dentatorubral-pallidoluysian atrophy: illustrative case. J Neurosurg Case Lessons. 2025 Jul 7;10(1):CASE2576. doi:10.3171/CASE2576. PMID: 40623331.

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