

## Quality of Life in Paediatric Survivors of Medulloblastoma

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### Abstract

Medulloblastoma (MB) is among the most common malignant paediatric brain tumours usually arising in the cerebellum. The treatment is surgical resection followed by craniospinal radiation with or without chemotherapy. We assessed the current literature on survivors of MB and their quality of life (QoL). The QoL of MB survivors is significantly compromised in terms of decreased neurocognitive functions, Intelligence Quotient (IQ), and social functioning. These also lead to a compromised overall performance, school performance, lack of employment, social isolation, and caregiver burden. The survivors often self-reported better performances as compared to objective performance and that rated by the caregivers. The predictors of worse QoL include earlier age at diagnosis, hydrocephalus, shunt placement, altered mental status at diagnosis, incomplete/subtotal resection of the tumour, and metastatic disease.

**Keywords:** Medulloblastoma, posterior fossa tumour, Quality of life, Paediatric brain tumours

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### Introduction

Childhood CNS tumours, while rare, surpass other cancers as the top reason for cancer mortality in those aged 0–14 years.<sup>1</sup> They are also the second most common childhood malignancy and the most common solid tumour in children.<sup>2</sup> Medulloblastoma (MB), is among the most common malignant paediatric brain tumours comprising up to 63% of intracranial embryonal tumours and 20% of all paediatric brain tumours.<sup>3</sup> The mainstay of therapy is maximum safe resection, followed by craniospinal radiation with or without chemotherapy. During tumour resection, gross total resection is superior to near-total resection in improving progression-free survival but there is no difference in overall survival, therefore surgical removal of small residual portions of MB is not recommended in cases where the chances of neurological morbidity are high. Nevertheless, maximal safe surgical resection remains the standard of care.<sup>2</sup>

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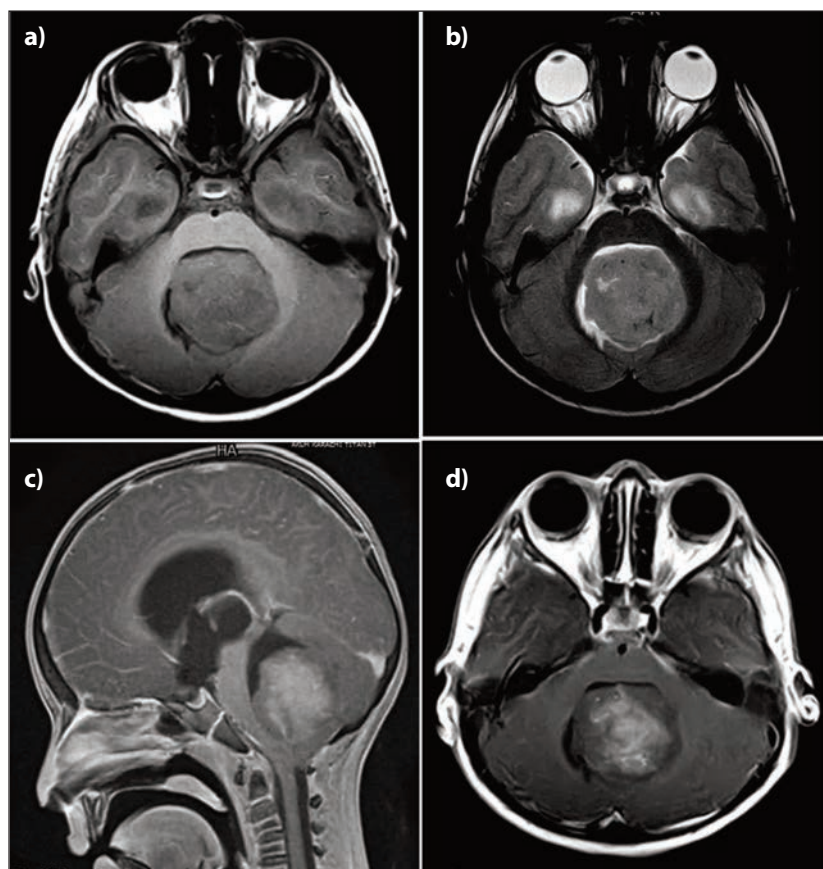
Conformal Radiotherapy improves survival in MB patients.<sup>2</sup> Patients with  $\geq 1.5$  cm<sup>2</sup> residual tumour on postoperative MRI are considered high risk and thus are considered for a higher amount of adjuvant craniospinal radiation although this may be associated with widespread adverse effects such as neuroendocrine dysfunction, growth disturbances, permanent neurocognitive disability, infertility, growth deformities, and secondary malignancies.<sup>4</sup>

### Review of Evidence

Packer et al., assessed IQ, school performances, and employment in 24 paediatric survivors of MB and reported a decreased overall performance for children which was significantly associated with altered mental status and oculomotor deficit at diagnosis. Decreased IQ was associated with age at the time of radiotherapy, the younger the child, the lower the IQ; mental status at diagnosis; the extent of resection with greater IQs in patients after total resection; and placement of shunt with greater IQs in unshunted patients. Hoppe-Hirsch et al., reported compromised QoL on a 10-year follow-up and reported that the psychological sequelae were significant and worsened over the years and also affected employment in later life.<sup>5</sup>

Ribi et al., assessed health-related QoL by assessing tumour control, neurological, endocrine, and neurocognitive complications and their impact on behavioural and psychological adjustments by using a semi-structured interview, Youth Self-report, Child Behaviour Checklist, and Paediatric Quality of Life inventory (PedsQL).<sup>6</sup> All patients had significant deficits in neurocognitive functioning with specific impairments in attention and processing speed, learning and memory, language, visual perception, and executive functions. Ribi et al., also reported that social functioning was rated by patients as the QoL dimension most affected. Parents of survivors rated their performance lower than those of the survivors themselves, implying that the survivors tend to underestimate their impairments.<sup>6</sup>

Maddrey et al., also reported a compromised overall quality of life as measured by QoL Index; along with psychologic functioning specifically IQ, attention, visuospatial memory, verbal memory; visuospatial abilities; and motor functioning. It was also stated that the survivors and their families do not report impaired QoL.<sup>7</sup> Kamran et al., reported a lower health-related QoL (as measured by



**Figure:** a) T1 axial image: Iso to hypointense lesion occupying the fourth ventricle, b) T2 axial image: Hyperintense lesion arising from the roof of 4th ventricle. c, d) T1 sagittal and axial images with contrast: Heterogeneous contrast uptake in the lesion and anterior displacement of brain stem due to mass effect. Ventriculomegaly can also be noticed.

PedsQL) in 50 patients following proton radiotherapy as compared to the normal population, however, the QoL seemed to improve over time in most patients except in survivors of high-risk MB and those with a shunt. The children with metastatic MB had scores that predictably declined with time.<sup>8</sup>

In a landmark paper de Medeiros et al., conducted a multinational survey including populations from North America, Europe, and East Asia and assessed 76 MB survivors who reported impairments in cognition and independent living in MB survivors, and concluded that the survivors do not interpret their abilities as burdensome compared to their caregivers. Western sites reported better performance compared to those from Eastern sites. The extent of resection had an impact on hearing abilities with those who underwent gross total resection having better scores for hearing.<sup>9</sup>

Contrary to the other reports, a study on QoL of paediatric patients following posterior fossa brain tumours out of which 30.6% were MBs in India reported a normal QoL, except in cases of large ventricle size, permanent hydrocephalus treatment, decreased family functioning, and lower family income.<sup>10</sup>

## Conclusion

The quality of life of MB survivors according to the current literature is significantly compromised in various aspects of psychosocial functioning. The predictors of a worse QoL included earlier age at diagnosis, hydrocephalus and shunt placement, altered mental status at diagnosis, incomplete/subtotal resection of the tumour, and metastatic disease.

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