Diffuse intrinsic pontine glioma in children.

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Background

DIPG (diffuse intrinsic pontine glioma) is one of the most lethal paediatric CNS malignancies, accounting for the bulk of deaths in this age range due to brain tumours [1].

Given this poor prognosis, novel treatment options are required to enhance these patients' prognosis and quality of life.

The majority of histologically confirmed DIPGs in children are High-Grade Gliomas (HGGs), mainly glioblastoma [2]. DIPG is frequently diagnosed based on clinical signs that are consistent with brainstem involvement in the context of typical MRI results. Despite the fact that the classic MRI signs of DIPG were described years ago [3], the characterization of unusual radiological features, especially when mild, is still debatable and subjective.

Since the early 1990s, when MRI was demonstrated to be a reliable diagnostic tool, biopsies have not been routinely taken unless in individuals with radiographically unusual tumours. Tumor biopsy has recently regained relevance in children with DIPG due to the minimal morbidity and mortality associated with this technique. Although molecular investigations of biopsy samples have helped us better understand the biology of DIPG [4], the need for a tumour biopsy is still debatable.

One risk with biopsies is that they may postpone the start of Radiation Therapy (RT) in individuals who are severely symptomatic. The paucity of clinically meaningful molecular markers to stratify therapy for children with DIPG, as well as the lack of promising medications to utilise based on this stratification, suggests that mandated biopsies may put patients at danger without providing personal benefit. Testing for the existence of the H3F3A K27M mutation, a poor prognosis marker [5], for example, is simple; however, in the absence of viable therapeutic alternatives, justifying a biopsy for the purpose of verifying a known mutation is problematic. The categorization of children based on molecular data provided at diagnosis in future clinical trials, especially once more potential targeted medicines become available, may allow for better therapy selection for affected children [6].

Previously, children with DIPG were treated in clinical trials similar to those used in adults with glioblastoma [7]. Unfortunately, no chemotherapy combination has been found to be effective in the treatment of DIPG. A study of 29 studies including 973 individuals treated between 1984 and 2005 found no evidence of a survival benefit with the inclusion of chemotherapy [7].

As a consequence of genome-wide studies of copy number anomalies, mutations, gene expression, and methylation patterns, our understanding of the molecular aspects of DIPG has advanced dramatically. DIPG has been shown to have clinical and molecular characteristics that distinguish it from other juvenile and adult HGGs [2]. This revolution in our understanding of molecular characteristics, particularly those considered to be actionable therapeutic targets, has resulted in a paradigm shift in the design of new clinical trials for children with DIPG, which now aim to investigate the cancers' unique biologic characteristics. The most promising present or future therapy methods for children with DIPG are as follows:

- Inhibitors of receptor kinase
- Essential cellular pathway inhibitors
- Mechanisms of DNA repair
- Epigenetic control
- Immunotherapy
- Modalities for delivering drugs in new ways

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***Received:** 06-Apr-2022, Manuscript No. AACIR-22-59808; **Editor assigned:** 08-Apr-2022, PreQC No. AACIR-22-59808(PQ); **Reviewed:** 16-Apr-2022, QC No. AACIR-22-59808; **Revised:** 20-Apr-2022, Manuscript No. AACIR-22-59808(R); **Published:** 27-Apr-2022, DOI:10.35841/56499-5.2.107*

***Citation:** Thapa V. Diffuse intrinsic pontine glioma in children. J Clin Immunol. 2022;5(2):107*