alterations in pLGGs are related to an activation of the mitogen-activated protein kinase (MAPK) pathway [5, 10]. This alteration can occur in presence of gene rearrangement, due to a tandem duplication at 7q34 determining a fusion between KIAA1549 and BRAF (KIAA1549-BRAF fusion) or by point mutation at the codon 600, which results in an amino acid substitution from valine (V) to glutamic acid (E) (BRAF V600E mutation) [10, 11]. KIAA1549-BRAF fusion is present in up to 66% of pilocytic astrocytomas and in 15% of all other low-grade gliomas, and it is recognized as a factor associated with a better prognosis [10, 12]. In addition, a further gene possibly involved in tumorigenesis is the one encoding the isocitrate dehydrogenase (IDH) enzyme, which catalyse the oxidative decarboxylation of isocitrate and therefore has a pivotal role in cell energy production. This gene has been indeed found to be mutated in gliomas [13]. When compared with adult lower-grade gliomas, IDH mutations are less frequent in the paediatric population particularly in younger children, and malignant progression is extremely rare in paediatric IDH wild-type LGGs [3].

Paediatric high-grade gliomas (pHGG), on the other hand, are one of the main causes of cancer-related death in children. These include various WHO grade 3 and 4 entities (some of them newly recognized in the WHO CNS5 under the group of paediatric-type diffuse high-grade gliomas), as well as diffuse midline glioma, H3K27-altered first introduced in the 2016 WHO classification [6]. The detection of H3K27 alteration, independent of the histological appearance (which could even be that of a low grade diffusely infiltrating lesion), constitutes classification as a WHO grade 4 [6]. Diffuse midline gliomas (DMG) can arise in any of the central nervous system mid-line structures (e.g. the brain stem, the thalamus, and the spinal cord). The H3K27 alteration is observed in up to 85% of diffuse intrinsic pontine gliomas (DIPG), which are aggressive malignant brainstem DMG for which the median survival is less than 1 year [14]. Of note, DIPG diagnosis can still be made based on clinical and imaging features alone, without tissue sampling. From the clinical point of view, these tumours manifest with a characteristic triad: multiple cranial neuropathies, long tract signs (hyperreflexia, clonus, increased tone, Babinski reflex), and ataxia. Classic features on MRI are a T1 hypointense and T2 hyperintense diffusely infiltrating lesion arising from and involving ≥ 50% of the pons [14].

Despite similar histological characteristics, pHGGs have different molecular features when compared with adult gliomas, both in terms of mutation pattern and in the prognostic implication of those mutations. In particular, approximately 40% of pHGGs are associated with tumour suppressor gene TP53 mutations [15, 16]. Compared with adults, pHGGs are less likely to bear epidermal growth factor receptor (EGFR) gene amplification and less likely to display mutations in the tumour suppressor PTEN [16, 17]. Up to one-third of hemispheric pHGGs carry mutations at position 34 (G34R/V) in H3F3A [3]. Conversely, hotspot mutations in IDH1/2 are rare in older adolescent and represent the lower age spectrum of adult gliomas [3, 18].

It is worth noting that familial syndrome might increase the occurrence of CNS paediatric tumours: in fact, 8–19% of these neoplasms is found in patients with genetic predisposition; this figure is significantly lower in adults [19]. These conditions include tuberous sclerosis, neurofibromatosis, Li-Fraumeni syndrome, rhabdoid tumour predisposition syndrome, von Hippel-Lindau disease, naevoid basal cell carcinoma syndrome, and Turcot's syndrome [20].

Clinical management of paediatric brain gliomas is challenging and requires a multidisciplinary approach in order to devise the best diagnostic and therapeutic strategies in each individual patient. Surgery represents the treatment of choice and can have the most relevant impact on patients' prognosis. If complete surgical resection is not feasible, biopsy or "debulking" can be considered, and adjuvant therapy with radiotherapy, chemotherapy, or a combination of both may be used. However, for pHGGs, due to possible persistence of microscopic disease, adjuvant therapy is proposed and performed even in case of complete tumour removal. In conclusion, the principal therapeutic approach in paediatric gliomas is represented by surgery in association with chemo- and/or radiotherapy [21].

Sensitive and effective non-invasive imaging procedures are especially needed in this "era" of new surgical techniques, radiotherapy planning, and novel systemic treatment. Although conventional MRI is the pivotal imaging procedure in paediatric brain gliomas to determine tumour location, presence of oedema, presence of intratumoural necrosis, cyst formation, haemorrhage, vascularization, mass effect, and contrast enhancement, it has some limitations in distinguishing tumour from tumour mimics and in defining tumour type and grade. Moreover, it does not always allow for precise delineation of tumour margins, distinguish cells in the tumour microenvironment, or inform about the metabolism or state of tumour cells.

Indeed, after treatment, the differentiation between true tumour remnants and treatment-related changes (e.g. "pseudoprogression" or "pseudoresponse") can be very difficult, specifically when an early response assessment is performed. Pseudoprogression occurs in approximately 21–44% of DIPG [22] and 7–12% of pHGG cases [23]; it is a subacute treatment-related reaction (occurring most frequently within the first 3 months after chemoradiotherapy) characterized by a "transient increase in tumour size due to an increase in blood–brain barrier permeability caused by treatment, resulting in increased oedema, or contrast enhancement, or both" [23].

Pseudoresponse is a different MRI phenomenon characterized by "radiographical improvement, with overall