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MYELOPROLIFERATIVE NEOPLASMS

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In addition to the chronic myeloid leukemia (CML) BCR-ABL1+, which will be discussed separately, classic myeloproliferative neoplasms (MPNs) include polycythemia vera (PV), essential thrombocythemia (ET) and primary myelofibrosis (PMF). These have a very low incidence in the pediatric age group, with about 0.82 cases for every 100 thousand patients, about 100 times less than in adults.[1,2]

Pediatric patients generally have a lower incidence of mutations commonly found in adults, thrombotic events and transformation to myelofibrosis and acute leukemia. There is no consensus on treatment in children, with little data in the literature.1-2 Most children with PV and ET are treated with supportive care and sometimes cytoreductive therapies.[3,4] Although extremely rare, PMF has a very heterogeneous phenotype in children, with variable evolution, from occasional spontaneous resolution to a rapidly progressive disease, sometimes fatal, curable only by HSCT.[5-7]

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