

Compilation of International PPB Registry and Literature Cases of Neoplasia and Dysplasia Associated with Cases of Pleuropulmonary Blastoma

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Associated Diseases ¹	Number of Affected Individuals	
	PPB Child	Family Members ²
Lungs		
PPB		7 families with 2 or more PPBs (other PPB-associated cysts in some of these families) 2 families with 1 PPB and 1 regressed PPB
PPB and familial lung cysts, not pathologically examined		5 families
Bilateral PPB	10	
PPB and contralateral lung cysts	13	
PPB and ipsilateral and contralateral cysts	4	
PPB and additional ipsilateral cysts	11	
PPB and second non-PPB malignant lung tumor	1	
Kidney		
cystic nephroma	8	7 (brother: 1)
cystic nephroma or renal cyst and small bowel polyps	5	
renal cysts, not biopsied	6	
Wilms tumor		1 (mother)
Intestinal polyps	7 ³	2
Thyroid – nodular hyperplasias, follicular and papillary carcinomas	8	10 families (many individuals incl. siblings & parents)
Mesenchymal proliferations		
bladder rhabdomyosarcoma	1	1
nasal chondromesenchymal hamartoma	3	
synovial sarcoma		1 (sister)
Blood Dyscrasias (including leukemia/lymphoma)	1	7 (mother: 1)
Gonadal tumors		
stromal sex-cord tumors, especially Sertoli Leydig tumors	3	4 (mother: 1)
germ-cell tumors, especially seminoma/dysgerminoma		7 (father: 1) ⁴
Brain tumors		
medulloblastoma	1	1
glioblastoma multiforme	1 ⁵	
ependymoma		2 (sister: 1)
primary brain sarcoma		1 (brother)
childhood brain tumor NOS		3
Miscellaneous		
cystic hepatic mesenchymal hamartoma	1	
bladder transition-cell carcinoma		1 (father) ⁶
bladder papilloma		1 (father) ⁷
globe melanosa		1
globe medulloepithelioma (dictyoma)		1 (father)
neuroblastoma		1 (sister)
anaplastic carcinoma of pharynx		1 (father) ⁸
Genetic Conditions		
neurofibromatosis type 1	2	

Notes:

¹ diseases listed occurred under age 20 years, unless otherwise noted

² first-degree relatives identified specifically

³ five of seven also had cystic nephroma

⁴ several of the gonadal tumors occurred in adults age 20 – 40 years

⁵ probably radiation-induced after PPB brain metastasis

⁶ age at diagnosis 37 years

⁷ age at diagnosis 33 years

⁸ age at diagnosis 13 years