Supplemental Table 1 Review of the literature

C	Reference[f]	Dicer1 Syndromic[b]	Dicer1 Non-syndromic[b]	Number of cases (n)[h]	Age [c]	Gender	Size (mm)[e]	Non-thyroid tumours	Histotype[g]	Aggressive features	Multifocality/ Bilaterality[d]	Encapsulation	Well-known somatic mutations
2008	1-(Post-therapy)	DICER1	NO	1	6	F	NA	PPB	FTC	recurrence, CI	NA	YES	NA
2008	2 –(Post-therapy)	DICER1	NO	1	16	F	NA	PPB, RMS,	miFTC and VA	CI	NA	YES	NA
2010	3[f]	NA	NA	1	46	F	20	SLCT	cystic PTC with oxyphillia	CI	NA	YES	NA
2010	4	DICER1	NO	3	34	F (2), M (1)	NA	CN, PPB	MNG [a]	NA	NA	NA	NA
2011	5	DICER1 (1)	Germline pathogenic mutation (n=6)	7	22 (syndromic cases)/ 9 (non-syndromic case)	NA	NA	SLCT, WT	MNG/cysyts [a]	NO	NA	NA	NA
2011	6	DICER1 (8)	Germline pathogenic mutation (n=31)	39	NA	NA	16 (only syndromic ones)	SLCT (8 syndromic cases)	NA	NO	NA	NA	NA
2012	7-Post-therapy)	NA	NA	1	9	F	27	PPB	FTC	CI	YES	YES	NO
2013	8	DICER1	NO	1	16	F	NA	SLCT, CN, PB, ME	PTC	NA	NA	NA	NA
2013	9	DICER1	NO	2	18	F	NA	SLCT	MNG	NA	YES	NA	NA
2014	10 [b]	DICER1	NO	1	9	M	30	CN	MNG	NO	YES	NA	NA

2014	11	DICER1	NO	1	5	F	NA	PPB, NCMH, CBME, ERMS, CN, SLCT	FV-PTC	NO	YES	YES	NA
2014	12	DICER1	NO	1	16	F	NA	WDFA, SLCT,	MNG	NO	YES	NA	NA
2014	13	DICER1	NO	2	14	F (1), M (1)	40	SLCT	MNG/ FA with PH	NO	YES	YES	NA
2014	14	DICER1 [1])	Germline pathogenic mutation (n=3)	4	32	F	NA	SLCT (1 syndromic case)	MNG	NA	YES	NA	NA
2014	15	DICER1 (3)	NO	3	8.3	F (2), M (1)	NA	PPB, cysyts, ME, cataract	FPTC (2), PTC	CI	NA	NA	NA
2014	16	DICER1 (5)	Germline pathogenic mutation (n=2)	7	27	F (3)/M (3)	NA	SLCT, CN, NCMH, PPB	PTC, DTC, MNG, Thyroid Cysts	NA	YES	NA	NA
2015	17	DICER1	NO	1	3	F	NA	SLCT	PTC	NA	NA	NA	NA
2016	18	DICER1 (2)	Germline pathogenic mutation (n=3)	5	11	F	NA	SLCT, CN	EFV-PTC (3), miFTC (mother)	NO	YES	YES	NO
2016	19	NO	Somatic mutation	4	NA	NA	NA	NO	miFTC and FA	CI	NA	NA	NO
2016	20	DICER1	NO	2	14	F	NA	OT, CP, L, CN, BCC, PM, ICT	MNG	NO	YES	NA	NA
2016	21	DICER1	NO	2	15	F	35	SLCT, CN, ERMS, yolk sac, pituitary blastoma	EFV-PTC (1) and wi-FTC (1)	CI, VI (both in FTC)	YES	YES	NA
2016	22	DICER1	NO	1	11	F	10	SLCT	HAN	NO	YES	YES	NA
2016	23	DICER1	NO	2	12	F/M	NA	Uterine myoma, PPB	Thyroid cysts	NA	NA	NA	NA
2017	24	DICER1	NO	1	12	F	20	ASK	mi FTC	CI	YES	YES	NA
2017	25	DICER1 (8Ma)	Germline pathogenic mutation (n=2Ma/12 Be)	22	16 (syndromic), 36 (non-syndromic)	M (1), F (9)	NA	PPB, pineoblastoma	microPTC(1), FV-PTC(3), miFTC(1), FTC(1), mix pattern PTC(1), macrofollicular PTC(1), multifocal PTC(1), classical PTC(1)	CI	YES	YES	NA
2017	26	DICER1	NO	1	27	F	NA	EAS	MNG	NA	NA	NA	NA
2017	27	NO	Germline pathogenic mutation (n=5 unaffected	5	23 (only 2 case	F	NA	NO	MNG [a]	NA	YES	NA	NA

			carrier)		available)								
2018	28	DICER1	NO	1	12	F	14	Cervix ERMS	WDTC-NOS	AI	YES	Partially	NO
2018	29	NO	Somatic mutation	1	45	F	28	NO	TCS	DM, AI, CI	NA	NA	NO
2018	30	NO	Germline pathogenic (n=4) and somatic mutation (n=5)	8	<18	F(7)/M(1)	27	NO	6 PTC (2 aggressive subtype), IFV-PTC, miFV-PTC (9) and 2 FND	ALL, CI, AI (1)	YES (1)	YES (2)	NO
2018	31	DICER1	NO	10	29	NA	NA	NCMH, SLCT, PPB, RMS, CN	DTC	NA	NA	NA	NA
2018	32	NO	Germline pathogenic mutation (n=10) somatic mutation (n=2)	10	14	F (7)/M (3)	T1	Na	NIFTP (3), FV-PTC (5), PDTC (2)	NO	YES (6)	YES (3)	TP53 (1)
2018	33	NO	Somatic mutation	1	11	NA	18	NO	SV- PTC	NO	NO	YES	NO
2018	34	DICER1	NO	1	58	F	NA	pituitary microprolactinoma	PTC	NA	NA	NA	NA
2018	35	DICER1	NO	1	13	F	70	SLCT, ERMS	FV-PTC	NA	YES	NA	NA
2018	36	DICER1 (1)	Germline pathogenic mutation (n=6), somatic mutation (n=3)	7	<18(6), 23(1)	F (5)/M (2)	NA	SLCT (1 syndromic case)	MNG	NO	YES	NA	NA
2019	37	NO	Germline pathogenic mutation	1	18	F	NA	NO	FV-PTC	NA	NA	NA	NA
2019	38	DICER1 (5)	Germline pathogenic mutation (n=5)	10	10.8 (syndromic cases)/ 15.4 (non-syndromic case)	F (7), M (3)	NA	SLCT, NB, cysts, ERMS	MNG	NO	5 of them bilateral/multifocal	NA	NA

Pleuropulmonary blastoma (PPB), Sertoli-Leydig cell tumour (SLCT), Embryonal rhabdomyosarcoma (ERMS), Wilms tumour(WT)nasal chondromesenchymal hamartoma (NCMH), medulla epithelioma (ME), Pineal blastoma (PB), ovarian teratoma (OT), colon polyps (CP), lymphoma (L), pilomatrixoma (PM), intracranial teratoma (ICT), endometrial adenosarcoma (EAS), anaplastic sarcoma of kidney (ASK), Well differentiated thyroid carcinoma (WDTC-NOS), thyroid carcinosarcoma (TCS), minimally invasive follicular thyroid carcinoma (miFTC),

follicular adenoma (FA), papillary thyroid carcinoma (PTC), invasive follicular variant papillary thyroid carcinoma (IFV-PTC), follicular nodular disease (FND), (NIFTP), poorly differentiated thyroid carcinoma (PDTC), Papillary hyperplasia (PH), multinodular goitre (MNG), vesicular adenoma (VA), encapsulated follicular variant of PTC (EFV-PTC), hyperplastic adenomatous nodule (HAN), solid variant of PTC (SV-PTC), widely invasive follicular thyroid carcinoma (wi-FTC), Angioinvasive (AI), capsule invasion (CI), distant metastasis (DM), vascular invasion (VI), acute lymphoblastic leukaemia (ALL). Malign (Ma), Benign (Be), Male (M), Female (F), NA: Non-available, NO: no

a-pathologically not proven (only radiologic finding)

b-The cases who has a family history, DICER1 germline alteration and at least 1 associated tumour previously reported in DICER1 syndrome were accepted as "syndromic". Carriers or non-carriers without exposing multiple tumours (only with thyroid disease) were accepted as non-syndromic.

c-For the case series, mean age was calculated and both for the case series and isolated cases, the onset accepted according to the first manifestation of thyroid disease.

d-MNG was accepted as multifocal disease due to its nature

e-Mean size was calculated for the case series. Age was not separated syndromic versus non-syndromic when calculated for mean.

f-We tried to exclude the overlapping cases in the different articles.

g-Some cases have two peaks for thyroid disease, only the onset disease was noted when available

h-Not only index or issued cases but also probands/relatives were included when mentioned in the articles of the table.

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