

# 시상하부 과오종에 대한 감마나이프방사선수술의 장기추적 결과 : 치료성적을 어떻게 높일 수 있을까?

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## Long-Term Follow-Up Results of Gamma Knife Radiosurgery for Hypothalamic Hamartoma : How can we Improve the Results?

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**Objective:** Hypothalamic hamartomas (HH) constitute rare developmental lesions associated with gelastic seizure, precocious puberty, and abnormal behavior. Treatment for this lesion is very complicated due to its location. Gamma Knife radiosurgery (GKS) may be an efficient and safe treatment option, which produces little morbidity. The authors evaluated the long - term results of GKS for the HH.

**Methods:** Eight patients with HH - related intractable gelastic seizure and/or precocious puberty underwent GKS between 1992 and 1996, with a mean age of 8.3 years at the time of GKS (range, 3.5 - 17.7 years). Three patients were presented with intractable gelastic seizure, and 4 patients with precocious puberty. One patient had both intractable seizure and precocious puberty. The mean follow - up duration was 76.6 (28.9 - 141) months.

**Results:** The mean marginal dose for the large sessile type HH with intractable gelastic seizure was 11.5Gy (range, 9 - 13Gy) and that for the small pedunculated type with precocious puberty was 27Gy (range, 20 - 34Gy). There was no lesion volume change on follow - up MRI. Intractable seizure disappeared in only 1 patient and was not improved in the other 3 patients. Precocious puberty was not resolved by GKS in all cases.

**Conclusion:** The long - term results of GKS for HH were not satisfactory. As for the control of epilepsy, the radiation dose in our cases was not enough to suppress and block the epileptogenic focus of HH and its propagation to surrounding tissue. Precocious puberty caused by HH is not indicated for GKS. If we could get better treatment outcome with higher radiation dose and/or new dose planning technique, GKS might be primary treatment option for HH.

**KEY WORDS:** Epilepsy · Gamma Knife radiosurgery · Gelastic seizure · Hypothalamic hamartoma · Precocious puberty.

### 서 론

(hypothalamic hamartoma) (ectopic) (tuber cinereum) (mammillary body) (precocious puberty) (seizure) 가 . 0.5~4cm (pedunculated type) (sessile type) 2)14)27)43)44)

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가 , (laughing noi-

ses) (clonic 대상 및 방법  
 facial grimace) (gelastic sei-  
 zure) 가 1992 9 1996 12 8  
 (cog-  
 nitive deterioration), (behavioral problems) Gamma Knife Unit Type B(Elekta AB, Stockholm,  
 가 Sweden) KULA system(ver-  
 (mental decline) 2)4)6)8)14)17)23)27)28)33)43)44) sion 5.4, Elekta AB, Stockholm, Sweden)  
 1934 MRI  
 100 가 3 가 5  
 43) 8.3(3.5~17.7)  
 가 Table 1  
 가 8)17)21)22)26)29)31)34)40)42) 4 , 1 2가  
 가 가  
 (luteinizing hormone, LH)  
 (prolactin) 가 가  
 (scalp EEG),  
 (ictal) (interictal)  
 1)3)5)7)16)20)21)29)36)41)43)45) (single photon emission computed tomo-  
 graphy, SPECT)  
 Engel  
 (Engel's classification)  
 76.6(28.9~141)  
 가 4 8 결과  
 간질발작의 특징과 뇌파소견

**Table 1.** Clinical features and radiological findings of hypothalamus hamartoma\*

No	Age (years)/Sex	Age at the seizure onset (months)	Symptom	Behavior abnormality	Mental retardation (IQ)	Type
1	11.4/male	20	GS, CPS	+	65	Sessile
2	15.6/male	30	GS, CPS, 2ndary GS	++	<50	Sessile
3	17.7/male	29	GS, CPS, PP	-	96	Sessile
4	4.2/female	6	GS, CPS, 2ndary GS	+	NA	Sessile
5	4.5/female		PP	-	NA	Sessile
6	4.2/female		PP	-	NA	Pedunculated
7	3.5/female		PP	-	NA	Pedunculated
8	5.7/female		PP	-	NA	Pedunculated

\*GS : gelastic seizure, CPS : complex partial seizure, PP : precocious puberty, 2ndary GS : secondary generalized seizure, NA : not available, - : none, + : mild, ++ : moderate

가 . 13.5(3~30) (slow  
 , background activity)  
 5.2(2~13) , (spike and wave complex)  
 (motor manifestation) (behavioral 4~5  
 arrest) (Table 1). (beta activity)가 (rhythmic build-

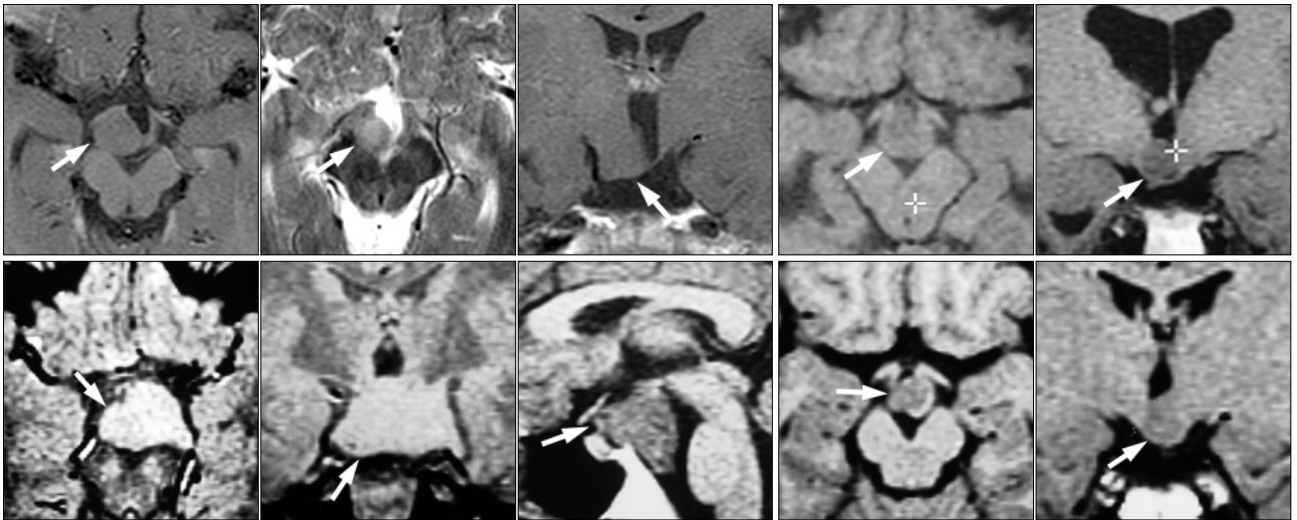


Fig. 1. MR images of sessile type hypothalamic hamartoma with intractable gelastic seizure (arrows : hypothalamic hamartoma).

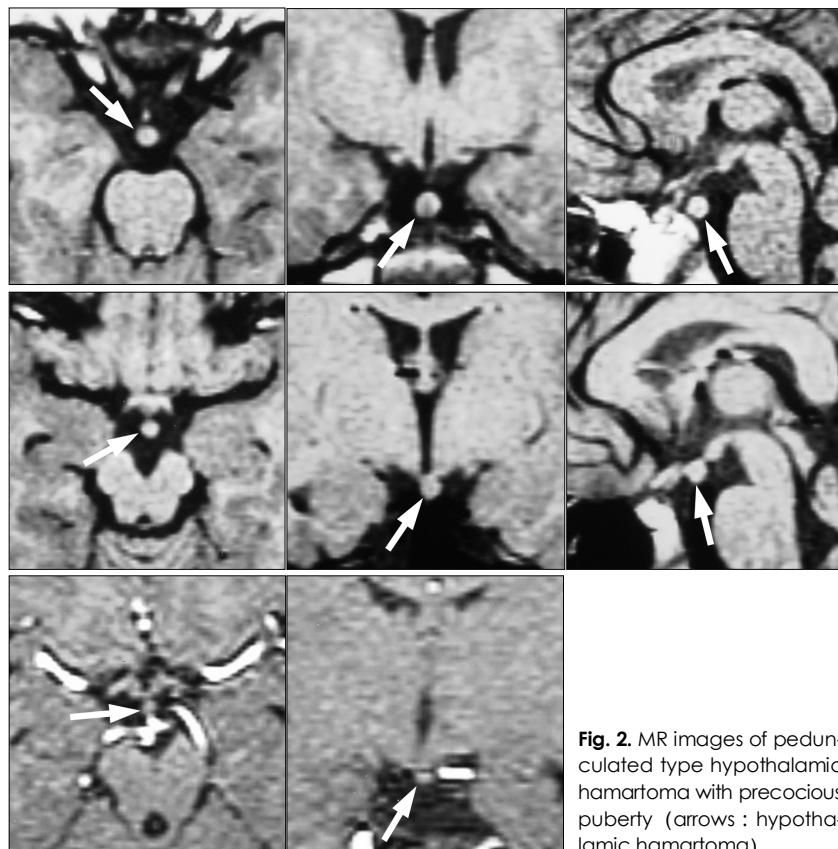


Fig. 2. MR images of pedunculated type hypothalamic hamartoma with precocious puberty (arrows : hypothalamic hamartoma).

up) , 7 , 18  
 1  
 3  
 MRI 소견과 임상 소견과의 관계 , (op-  
 MRI tic neuropathy)  
 고 찰  
 (Fig. 1, 2).  
 가 ( 0.12cm<sup>3</sup>, 0.09~0.15  
 cm<sup>3</sup>) 가 , (gray matter)  
 ( 27Gy, (hyperplastic neuronal tissue)  
 20~34Gy), 2)14)27)43)44)  
 가 ( 3.66cm<sup>3</sup>, 1.17~9.13  
 cm<sup>3</sup>) , 가  
 ( 11.5Gy, 9~13Gy). 가  
 10Gy (Table 2).  
 치료 후 경과 및 치료성적 , 가  
 10 가 ,  
 Table 2 , (drop attack)  
 (central necrosis)가 (secondary generalized epilepsy)  
 가 4)6)8)17)23)27)28)33)  
 72.4 1 2)8)9)13)35)42)  
 2 , 가  
 63.1 130 (lesionectomy)  
 (endoscopic disconnection) (Table 2).

**Table 2.** Radiosurgical and follow-up data

No	Volume (cc)	Maximal dose (Gy)	Marginal dose (Gy)	Dose to optic pathway (Gy)	Change of tumor volume	Follow-up duration (mo)	Outcome	Treatment after GKS*
1	1.17	23	11.5	<8.0	No change	33.1	Engel's Class IV	Anticonvulsants
2	2.91	18	9	<9.0	No change	130.0	Engel's Class III	Endoscopic disconnection
3	9.13	25	12.5	<9.5	Minimal central necrosis	72.4	Engel's Class I, no response for PP	Hormone therapy
4	1.44	26	13	<7.8	Minimal central necrosis	63.1	Engel's Class IV	Endoscopic disconnection
5	0.095	40	20	<6.4	No change	141.0	Partial response**	Hormone therapy
6	0.14	68	34	<8.4	No change	54.2	Partial response***	Hormone therapy
7	0.087	60	30	<9.0	No change	89.9	No response	Hormone therapy
8	0.15	40	24	<6.8	No change	28.9	No response	Hormone therapy

\*GKS : Gamma Knife radiosurgery, \*\* : initial response for 7 months and then progressed, \*\*\* : initial response for 18 months and then progressed

6)8)17)21)22)26)29)31)34)40)  
 10Gy 20Gy , 10)24)25)  
 4)6)8)17)23)28)33) 10~15Gy , 45) , Regis 36)  
 2)14)15)21)27)44) 가  
 17Gy  
 가 가 (resectability) , 13Gy  
 가 17Gy  
 가 2)9)14)44) 4 , 가 가 4 1  
 4 가 , 가 11.5(9~13)Gy 13Gy ,  
 Regis 가  
 1)17)29)31)33)39)  
 가 , 가  
 가 가  
 가 가 , 가  
 가 가 , 가  
 가 , 가  
 가 가 , 가  
 가 , 가  
 가 , 가  
 가 (subthalamus)  
 가 , 가  
 8~10Gy 가  
 가  
 (amygdala) (hippocampus), 12)20)37) 38)  
 (epileptogenesis) 30)



결 론

가 ,

17~18Gy

가

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