

CHARGE증후군 1예

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A Case of CHARGE Syndrome

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-ABSTRACT-

Bilateral congenital choanal atresia, one of the most common nasal anomalies, is in need of emergency treatment in the otolaryngologic field. CHARGE syndrome is a very rare disease with six congenital malformations which are characterized by coloboma of the eye, hearing deficit, choanal atresia, retardation of growth, genital defect and endocardial cushion defect. We report a case of CHARGE syndrome that is composed with bilateral choanal atresia, hearing deficit, coloboma, patent ductus arteriosus and other anomaly such as hiatal hernia with reviewing reported references. (J Clinical Otolaryngol 2003;14:137-140)

KEY WORDS : Choanal atresia · CHARGE syndrome.

서 론

증 례

1-4) CHARGE 3,550 gm 41 , 가
(Apgar score) 1 /5 8 9 .
3) CHARGE , , , . 가
(coloboma), , , 86 .
(nasoga-
CHARGE 1 stric tube)
McGovern nipple

: 2002 12 31

: 2003 4 6

: , 570 - 711

344 - 2

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(Fig. 1).

4 mm

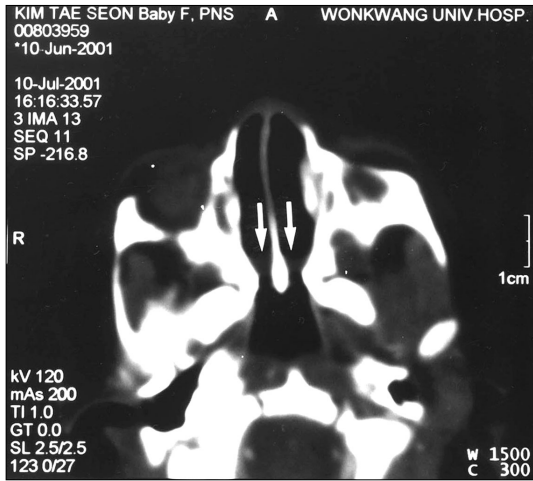


Fig. 1. Bilateral membranous choanal atresia in an infant. Axial CT shows soft-tissue membranes (arrows) extending from the lateral aspects of the vomer to the medial and posterior maxilla. The width of the vomer and the diameter of the posterior choanae are normal in this patient.

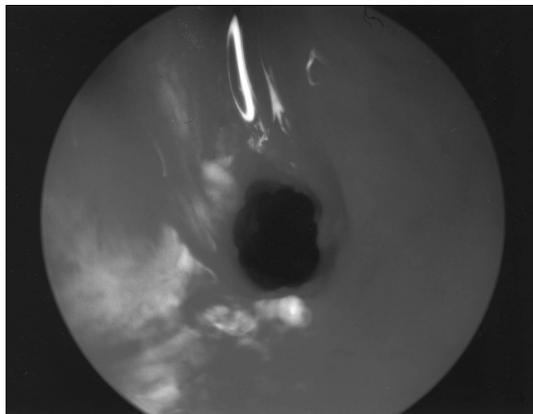


Fig. 2. Intraoperative picture of a newly formed right-sided choanae.

Nd : YAG
(Fig. 2), 4 mm
(circum-septal suture)

(patent ductus arterio-
sus)
가 (microphthalmia)
10 mm 6 mm
(coloboma)
가 가
가
(hiatal hernia)
6
4 Nd :
YAG
8
6

고 찰

5,000 10,000
가 , 가
10~15% 5)
CHARGE Coloboma, Hearing deficit, Atr-
esia choanae, Retardation of growth, Genital defect,
Endocardial cushion defect 6
가 6)
4가 가
(polydactyly), - - (nasal - au-
ricular and palatal deformities), Crouzon ,
(craniosynostosis), (microenceph-
aly), (meningocele), (meningoence-
phalocele), ,
(hypertelorism) 7)
CHARGE
10~15%, 6) 56% 8)
, Edward , 2) (chondrodys-

: CHARGE 1

plasia punctata)⁴⁾

¹²⁻¹⁵⁾

(urethral sound), mi-

2 : 1 가 , 70%가

crorongeur, ,

CO2, Nd : YAG, KTP,

가 holmium : YAG . Healy ¹⁶⁾

90%, 10%

CO2

30%, - 70%

가 1 mm가 mic-

rorongeur .

⁹⁾ 가

가 가 가

(buccopharyngeal membra-ne)

가 ,

³⁻¹²
¹⁻³⁾¹⁵⁾ Prasad ¹⁷⁾

(nasobuccal membra-ne)

mitomycin

¹⁰⁾

CHARGE

가

4 mm Nd : YAG

³⁾ - (crani-

6

ofacial abnormality)

4

8

6

⁶⁾

가

CHARGE

1

70%

CHARGE

6가

, 1

가

⁸⁾

⁸⁾

CHARGE

6가

Falot 4

가

4가

CHARGE

¹¹⁾

CHARGE

1

중심 단어 :

. CHARGE .

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