



Endoscopic dacryocystorhinostomy as an option management of syndrome-related congenital dacryocystocele: a case report

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ABSTRACT

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Congenital dacryocystocele (CD) is a rare anomaly of the medial region of the orbit, caused by distal at the level of the valve of Hasner and proximal at the level of the valve of Rosenmuller obstruction of the lacrimal system. It may present as isolated abnormalities or maybe associated with syndromes. We described a neonate with a history of bilateral enlarged lacrimal sacs below medial canthal tendon with blue-grayish color and epiphora. Computed tomography (CT) scan resulted in congenital dacryocystocele. She was given topical antibiotics and Crigler massage as conservative treatment. In the first week, the right eye got a complete resolution. Otherwise, the left eye's lesion was worsening. The patient underwent endoscopic dacryocystorhinostomy (En-DCR) and silicone intubation on the left eye. Epiphora resolved with no recurrence at one month follow up. Conservative treatment and En-DCR have a success rate with no complication and widely used nowadays. In the case of CD associated with a syndrome, multidiscipline workups and treatment are essential.

ABSTRAK

Dakriosistokel kongenital adalah suatu kelainan yang jarang dijumpai, terjadi pada bagian medial dari orbit yang disebabkan oleh obstruksi sistem lakrimal pada bagian distal setinggi katup Hasner dan proksimal setinggi katup Rosenmuller. Dakriosistokel kongenital dapat terjadi sebagai kejadian tunggal diubah menjadi dapat berdiri sendiri atau berhubungan dengan suatu sindrom. Pada kasus ini akan diperlihatkan suatu kondisi neonatus dengan riwayat pembesaran bilateral pada daerah sakus lakrimal di bagian bawah tendon kantung medial yang menjadi warna kebiruan disertai dengan gejala epifora. Hasil dari *computed tomography* (CT) scan menunjukkan kondisi dakriosistokel kongenital. Pasien diberikan terapi secara konservatif dengan antibiotik topikal dan pijat Crigler. Pada minggu pertama, mata kanan menjadi sembuh total namun pada mata kiri kondisi memburuk. Pasien dilakukan *endoscopic dacryocystorhinostomy* (En-DCR) dan pemasangan *silicone tube* pada mata kiri. Pada bulan pertama kontrol, gejala epifora membaik dan tidak terjadi rekurensi kondisi. Melalui hal ini dapat dilihat bahwa terapi konservatif dan En-DCR memiliki angka kesuksesan baik tanpa ditemukan komplikasi sehingga digunakan sebagai pilihan terapi saat ini. Pada kasus ini dengan kecurigaan disertai dengan sindrom, maka pemeriksaan dan tatalaksana secara multidisiplin merupakan suatu hal yang penting.

Keywords:

congenital dacryocystocele
epiphora
endoscopic dacryocystorhinostomy
syndrome

INTRODUCTION

Dacryocystocele is a persistent membrane at the level of the Hasner valve and functional obstruction at the level of Rosenmuller valve or common canaliculus followed by the dilation of the lacrimal sac. It is called mucocele when its contents of mucus and amniocoele, that contents amniotic fluid.^{1,2} The dilation is caused by the excretion of intraluminal goblet cells which are accumulated and trapped within the lacrimal sac with the appearance of the bluish lesion over the lacrimal sac.³ One of the classifications is congenital dacryocystocele (CD), which may present directly after birth or within four weeks after. In some cases extend intra-nasally as cysts and causes a respiratory obstruction that needs urgent treatment.^{4,5} The diagnosis of dacryocystocele could be based on clinical features with the feature of cystic swelling which is located inferior to the medial canthus. However, in some cases, periorbital ultrasonography, endoscopic nasal examination, CT, or MRI imaging plays a role in diagnosis and excluding the differential diagnosis. The similar clinical features are found in capillary haemangioma, meningoencephalocele, mucocele, encephalocele, and nasal glioma.^{2,3}

The management of CD is still controversial. Schanall and Christian⁵ found around 76% of CD resolved spontaneously within 2 weeks with conservative treatment such as warm compresses, massage, prophylactic topical and systemic antibiotic. Whereas Becker found 37.9% of CD developed dacryocystitis that recommended for early surgical treatments and case with recurrent epiphora, pyocele, fistula formation, intranasal cyst, lacrimal sac abscess, cellulitis, and a respiratory distress.^{1,6,7} If the lacrimal sac worsens after two weeks of massage, it will be treated with probing and if fails, we consider surgical interventions such as endoscopic marsupialization, endoscopic intranasal dacryocystorhinostomy combined with marsupialization, or endoscopic

intranasal dacryocystorhinostomy or nasolacrimal duct surgery. The study from Zhang *et al.*² reported that all three previous procedures (endoscopic marsupialization, endoscopic intranasal dacryocystorhinostomy combined with marsupialization, or endoscopic intranasal dacryocystorhinostomy or nasolacrimal duct surgery) healed after one year of follow-up with no recurrence of symptoms and sign moreover none of the children needed re-operation. However, those procedures need an experienced surgeon for the accuracy of surgical positioning to ensure there is no damage on the mucous membrane of lacrimal duct system outside the operation area. It will prevent the formation of false duct, infection of lacrimal duct, pain, and postoperative nasal bleeding.²

CASE

A 2-week-old girl patient showed a bluish lesion on the medial side of both eyes. It is followed by watery eyes and serous discharge (FIGURE 1A). She was delivered at the local hospital with palatoschisis, polydactyly, anorectal malformation, and dacrocystocele. It was suggested that the patient had Chromosome 2q35 Syndrome. The previous ophthalmologist prescribed conservative treatment with antibiotic eye drops. One week later, the lesion on the right eye had complete resolution. However, the lesion on her left eye became worse and bigger in two weeks. A topical antibiotic was continued with the addition of artificial tears eye drop four times a day for both eyes and Crigler massage daily by her parents.

At the age of eight week, the mass on the left eye was enlarged to a size of 2cm x 2cm, with firm consistency, without any discharge (FIGURE 1B). CT scan with contrast was conducted and was suggestive of dacryocystocele, a hypodense lesion on medial canthal region that connects with the nasolacrimal duct, and no erosion or bone destruction (FIGURE 2).

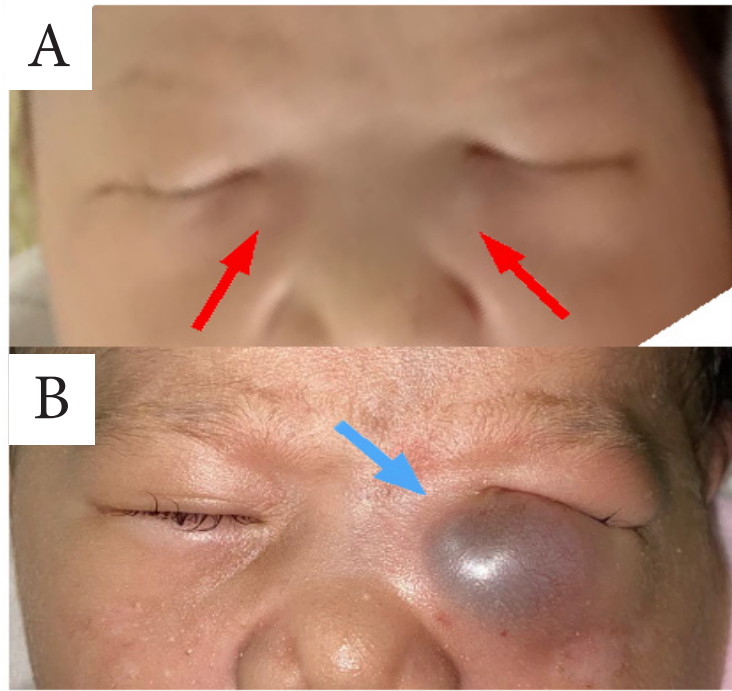


FIGURE 1. Dacryocystocele Developments. A). At 2-week-old girl, first time symptom appeared bluish lesion (black arrows) in the area of both lacrimal sac which is followed by watery eyes and serous discharge; B). At 8 weeks old, the mass on the left eye (red arrow) became bigger with the measurement 2 cm x 2 cm, firm in consistency, without any discharge

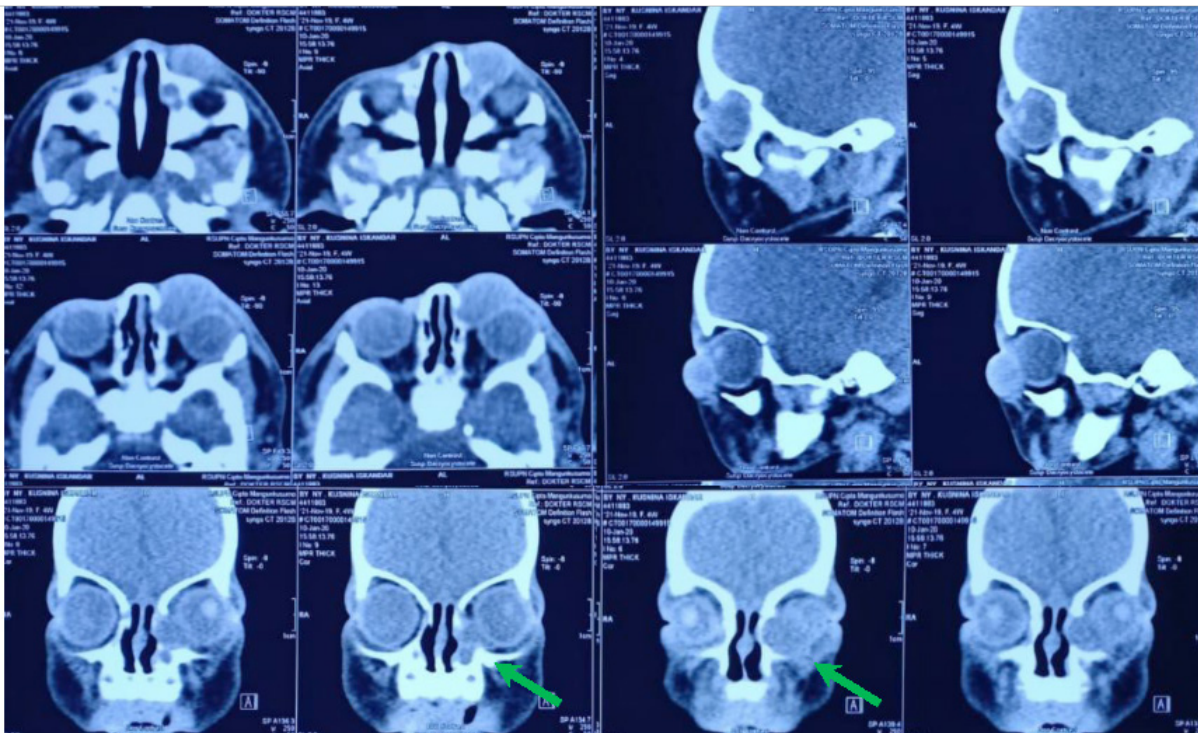


FIGURE 2. Orbital CT scan with contrast of the patient. There is hypodense lesion on medial canthal region that connects with nasolacrimal duct (green arrows)

The En-DCR (FIGURE 3) opted since there is no resolution of the left eye. It is followed by nasal mucosal flap in situ at the level of the middle turbinate and the pus came out and collected for culture examination. The procedure continued with an insertion of introducer probe through the inferior lacrimal punctum to the lacrimal sac and nasal cavity. An irrigation test was performed to examine lacrimal canaliculi patency and the result was positive. The insertion of a silicone tube was performed in both superior and inferior lacrimal punctum to the nasal cavity and followed by surgical knots on

the distal end of the silicone tube.

The patient was discharged with a post-operation treatment include prednisolone acetate and levofloxacin eye drop six times daily, amoxicillin syrup, ibuprofen, and advice of regular follow up. One month post-operation there is a good resolution of the lesion, no new complaint nor complication, and resolution was achieved. Three months post operation, the patient was still in a good condition without any complication and the removal of silicone tube was done (FIGURE 4).

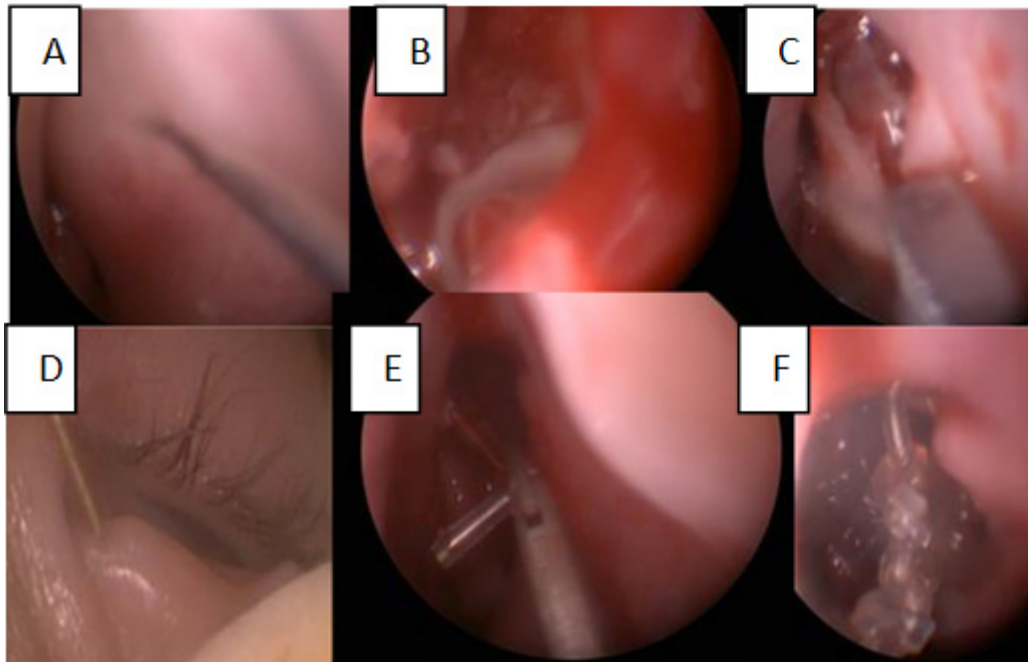


FIGURE 3. Intraoperative Pictures. (A) Creating nasal mucosal flap using sickle knife at the level of medial canthus; (B) Pus came out of the mass; (C) Incision at the level of media nasal concha to visualize lacrimal sac; (D) Introducer probe inserted through the inferior lacrimal punctum; (E) A silicone tube was then inserted; (F) Surgical knots were made on the distal end of silicone tube

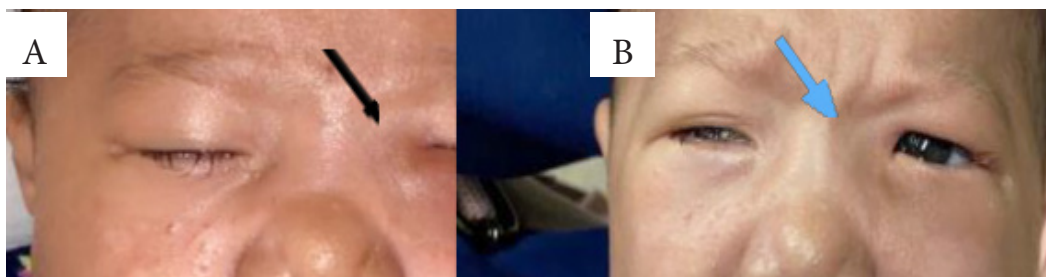


FIGURE 4. Post-operation Picture. (A) One day post-operation. Minimal edema at inferior palpebra (black arrow), (B) One month post-operation. Complete resolution seen on the left eye (blue arrow)

DISCUSSION

Congenital dacryocystocele is a rare anomaly of a newborn. The incidence of case is 0.08 to 0.1% and 25% are bilateral.⁷ Among them, they are developed the disease within 1-14 days after birth and predominantly female.² They may present as isolated abnormalities or maybe associated with syndromes, such as brachio-ocula facial syndrome, down syndrome, lacrimo-auriculo-dento-digital syndrome, chromosome 2q35 syndrome.^{1,3} In our case, the patient was female with a bilateral bluish mass that appears when she was 2 weeks old with the other abnormalities include palatoschisis, polydactyly, and anorectal malformation. These findings were important to be looked for any associated syndromes.

When the diagnosis of CD is uncertain, the radiology workup such as CT scan, magnetic resonance imaging (MRI), or ultrasonography (USG) is useful. The CT scan provides good detail of showing the bony details with CD's classic triad of cystic medial canthus masses, nasolacrimal duct dilatation, and contiguous submucosal nasal cavity mass in inferior meatus, formed by the ballooning of inferior imperforate membrane into the nasal cavity. The CT can also determine the diagnosis of lacrimal cysts, excluding brain encephalocele, sweat gland cysts, dermoid cysts, or other lacrimal abnormalities. MRI is good at cyst contents visualization and meningoencephalocele identification, USG gives information about the lacrimal sac and surrounding structures.^{6,8-10} In this case, a CT scan was chosen since the patient had lacrimal abnormalities and midfacial bone structure anomalies. The patient's result of CT scan was a hypodense lesion on the medial canthal region that connects with the nasolacrimal duct and suggestive of a dacryocystocele.

The initial treatments of CD are

Crigler massage, warm compress, topical and systemic antibiotics. If there is no resolution within two weeks, lacrimal probing is the next step for small lesions and spacious nasal passages, moreover, surgical intervention using En-DCR and/or intranasal marsupialization are chosen for cases with dacryocystitis or progressively worsening.¹¹⁻¹³

In this case, we performed the En-DCR technique since the lesion was huge and no resolution with conservative treatment; moreover we could directly visualize the CD from the nasal cavity. Probing and marsupialization was not done since it could be a false route in this patient. Before the procedure, it is mandatory to ensure that the procedure is feasible by the complete nasal and facial examination. During operation, it is imperative to ensure that sedation will not compromise the airway. The En-DCR procedure consists of four parts (1) fashioning of a nasal mucosal flap, (2) osteotomy, (3) opening of the lacrimal sac and the creation flaps, (4) manipulation and replacement of flap with the placement of the silicone stent.^{14,15} Similar to other literature, En-DCR is preferred for young patients, high-risk keloid scar, and female advantages since there are no external scar, less bleeding, less operation time, better visualization of the nasal area, and no medial canthal wound which maintain good blink reflex.^{14,16} In our case, there was no intraoperative or postoperative complication with no more bluish lesion, no more epiphora, and a the silicone tube is still in place. A study from Sarbjana *et al*,¹⁸ uses the following postoperative conditions without epiphora, normal tear meniscus height, or complete clearance of 5 min dye after instilling 2% fluorescein drop solution into the inferior conjunctival fornix as one of the criteria of a good surgery outcome.

As our case of CD was suggestive of a syndrome, we consulted the patient to pediatrician genomic experts for

further evaluation. It was suggestive of chromosome 2q35 syndrome since the patient has clinical findings of polydactyly, palatoschisis, and dacryocystocele. Chromosome 2q35 Syndrome is a very rare congenital syndrome related to other diseases and abnormalities. The most common abnormalities are syndactyly, but the other abnormalities which are known to be related are polydactyly, palatoschisis, ectodermal dysplasia, cryptophtalmos, craniosynostosis, dacryocystocele.^{1,19} Nevertheless, genetic testing should be performed to confirm a definite diagnosis of this patient. By the follow-up evaluation, the test has not been done due to the parents' patient refusal.

CONCLUSION

Congenital Dacryocystocele (CD) is a rare congenital anomaly, as an isolated or syndrome condition. Diagnosis of CD is based on clinical features. Computed tomography scan or MRI can be used to confirm this condition. Conservative treatment with massage, topical and systemic antibiotics are the first line of therapy and surgical intervention should be considered if there is no resolution. En-DCR was successful in the treatment of congenital dacryocystocele in this case. In the case of neonates with a syndrome, multidiscipline workup and treatment are essential.

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REFERENCES

1. Lueder GT. The association of neonatal dacryocystoceles and infantile dacryocystocele with nasolacrimal duct cysts. *Trans Am Ophthalmol Soc* 2012; 110:74-93.

2. Zhang Y, Fan Y, Fan J, Cui Y. Selection of surgical intervention for congenital dacryocystocele. *Eur J Ophthalmol* 2019; 29(2):158-64. <https://doi.org/10.1177/1120672118785276>
3. Davies R, Watkins WJ, Kotecha S, Watts P. The presentation, clinical features, complications, and treatment of congenital dacryocystocele. *Eye* 2018; 32(3):522-6. <https://doi.org/10.1038/eye.2017.235>
4. Foster JA, Carter KD, Durariraj VD, Kavanag MC, Korn BS, Nelson CC, *et al*. Basic and Clinical Science Course (BCSC), Section 7: Orbit, Eyelids and Lacrimal System. San Fransisco: American Academy of Ophthalmology; 2015.
5. Schnall BM, Christian CJ. Conservative treatment of congenital dacryocystocele. *J Pediatr Ophthalmol Strabismus* 1996; 33(5):219-22.
6. Becker BB. The treatment of congenital dacryocystocele. *Am J Ophthalmol* 2006; 142(5):835-8. <https://doi.org/10.1016/j.ajo.2006.05.043>
7. Thakrar P, Chivate R, Choudhary R, Patkar D, Deshmukh S. Congenital Bilateral Dacryocystocele. *Int J Heal Sci Res* 2016; 6(3):329-35.
8. Ali MJ, Psaltis AJ, Brunworth J, Naik MN, Wormald PJ. Congenital dacryocystocele with large intranasal cyst: Efficacy of cruceate marsupialization, adjunctive procedures, and outcomes. *Ophthalmic Plast Reconstr Surg* 2014; 30(4):346-51. <https://doi.org/10.1097/IOP.0000000000000206>
9. Pujari A. Congenital dacryocystocele. *BMJ Case Rep* 2016; 2016:bcr2016218029. <https://doi.org/10.1136/bcr-2016-218029>
10. Lorena SHT, Gonçalves ED, Silva JAF. Congenital dacryocystocele: Case report and treatment. *Rev Bras Oftalmol* 2014; 73(4):243-5.
11. Raslan OA, Ozturk A, Pham N, Chang J, Strong EB, Bobinski M. A Comprehensive Review of

- Cross-Sectional Imaging of the Nasolacrimal Drainage Apparatus: What Radiologists Need to Know. *Am J Roentgenol* 2019; 213(6):1331-40. <https://doi.org/10.2214/AJR.19.2150>
12. Mirza AA, Alsharif AF, Elmays OA, Marglani OA. Foreign body mimicking malignancy in acquired dacryocystocele. *Clin Case Reports* 2017; 5(3):296-9. <https://doi.org/10.1002/ccr3.826>
 13. Gendeh BS. Endoscopic dacryocystorhinostomy. In *Tech Open* 2018; 7(2):153-9. <https://doi.org/10.5772/intechopen.81831>
 14. Leatherbarrow B. *Oculoplastic Surgery*. 3rd Edition. Thieme Medical Publishers; 2020.
 15. Al-Qahtani AS. Primary endoscopic dacryocystorhinostomy with or without silicone tubing: a prospective randomized study. *Am J Rhinol Allergy* 2012; 26(4):332-4. <https://doi.org/10.2500/ajra.2012.26.3789>
 16. Saeed BMN. Endoscopic DCR without stents: Clinical guidelines and procedure. *Eur Arch Otorhinolaryngol* 2012; 269(2):545-9. <https://doi.org/10.1007/s00405-011-1727-3>
 17. Celenk F, Mumbuc S, Durucu C, Karatas ZA, Aytaç I, Baysal E, et al. Pediatric endonasal endoscopic dacryocystorhinostomy. *Int J Pediatr Otorhinolaryngol* 2013; 77(8):1259-62. <https://doi.org/10.1016/j.ijporl.2013.05.004>
 18. Sarbajna T, Takahashi Y, Regina M, Valencia P, Ana-magadia MG, Ishikawa E, et al. Dacryoendoscopy-assisted nasal endoscopic marsupialization for congenital dacryocystocele. *Int J Pediatr Otorhinolaryngol* 2018; 115:54-7. <https://doi.org/10.1016/j.ijporl.2018.09.017>
 19. Malacards. Human disease data base. 2020. Available at: https://www.malacards.org/card/chromosome_2q35_duplication_syndrome#: