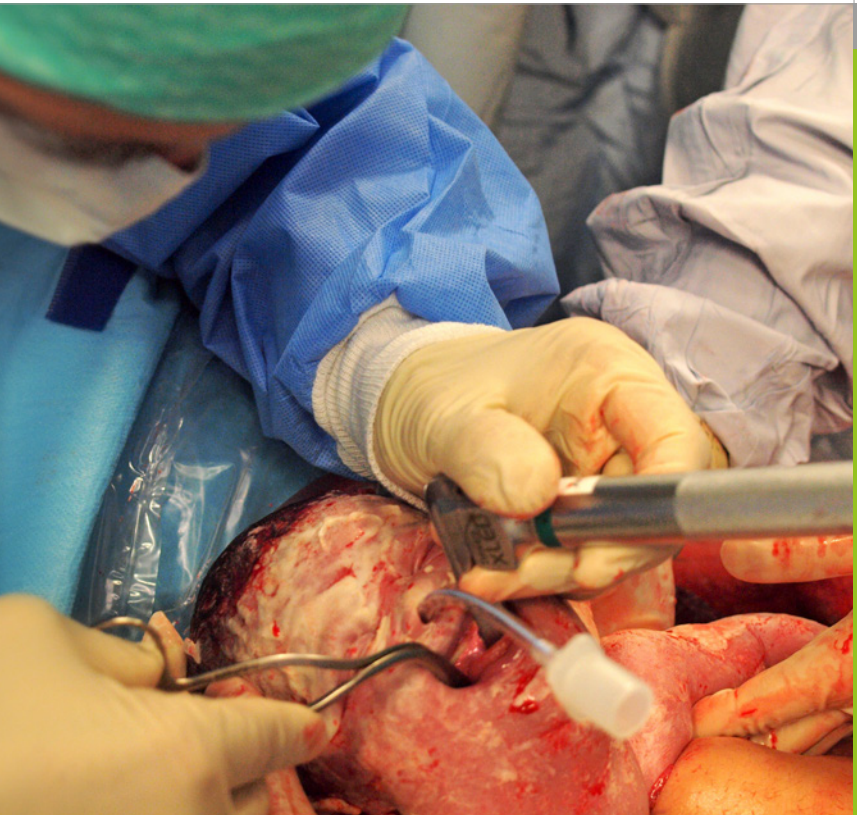
SWISS SOCIETY OF NEONATOLOGY

EXIT procedure in a patient with a severe lymphangioma colli



April 2016

Gubler D, Jöhr M, Winiker H, Hodel M, Berger TM,
Neonatal and Pediatric Intensive Care Unit (GD, BTM),
Pediatric Anesthesiology (JM), Pediatric Surgery (WH),
Children's Hospital of Lucerne, Department of Fetal
Medicine, Neue Frauenklinik Lucerne (HM), Switzerland

Lymphatic malformations occur in 1:2'000 – 4'000 live births (1). Lymphangioma colli is a relatively uncommon malformation that can cause a variety of complications depending on its precise location, type and size. Cervical macrocystic lymphatic malformations are also called cystic hygromas. Predominantly, they are located in the neck and axillary regions (95 %) and are found in the skin, mucosa, soft tissue and rarely in internal organs (2, 3).

Lymphangiomas are mostly congenital malformations resulting from erroneous embryogenesis (4). They can also develop after lymphatic obstruction, inflammation or trauma. These acquired forms are much less common in children and are not discussed further in this article.

Infants with prenatally diagnosed mass lesions in the area of the upper respiratory tract may be at risk to develop potentially life-threatening airway obstruction after delivery. High quality fetal imaging studies allow to better define this risk. In severe cases, an EXIT (ex utero intrapartum treatment) may be a suitable option to provide sufficient time to secure the airway before separating the infant from the placental circulation.

At 22 weeks of pregnancy, routine fetal ultrasound examination revealed a cystic mass in the area of the fetal neck of this female infant (Fig. 1). The findings were felt to be consistent either with a lymphangioma colli or a teratoma. Over the following weeks, the mass continued to increase in size (Fig. 2) and pregnancy was complicated by polyhydramnios most likely secondary to impaired fetal swallowing. Given the impressive size of the lesion and its location, a multidisciplinary team from fetal medicine, neonatology, pediatric otolaryngology, surgery and anesthesiology decided to propose an EXIT procedure. The parents agreed, and, after meticulous planning, the infant was delivered by elective Caesarean section at 37 5/7 weeks of gestation.

Following uterotomy, the head and the left arm were brought to the surface while the umbilical cord remained attached to the placenta. The vocal cords could be visualized by direct laryngoscopy without difficulty and the infant was intubated with a 3.5 endotracheal tube (Fig. 3, 4). Once the airway was secured, the female baby was fully delivered and the umbilical cord was cut seven minutes after uterotomy.

The extent of the neck mass was further examined with ultrasound and MRI (Fig. 5). Postnatal histopathology confirmed polycystic lymphangioma colli without signs of malignancy. On day eight of life, the

larger cysts were aspirated, followed by instillation of OK-432 (Picibanil®). Although the procedure was repeated two weeks later, no significant reduction of the mass was achieved. Therefore, extensive surgical excision was performed at seven weeks of age. At the same time, a tracheostomy was done. One month later, a gastrostomy tube was placed. Soon thereafter, the girl was discharged from hospital and followed on an outpatient basis.

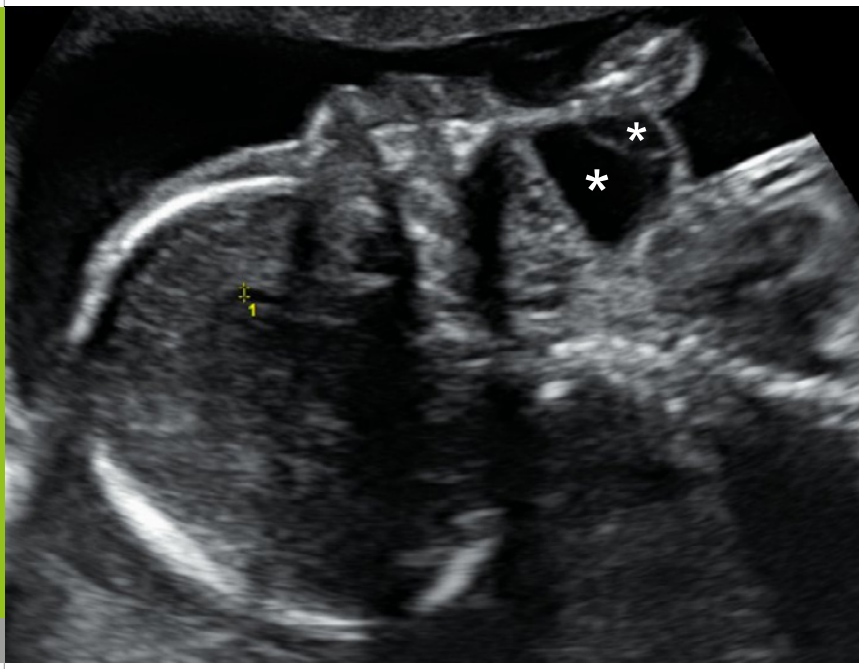


Fig. 1

Fetal ultrasound examination at 22 weeks of gestation: a cystic mass in the neck region is demonstrated (asterisks).

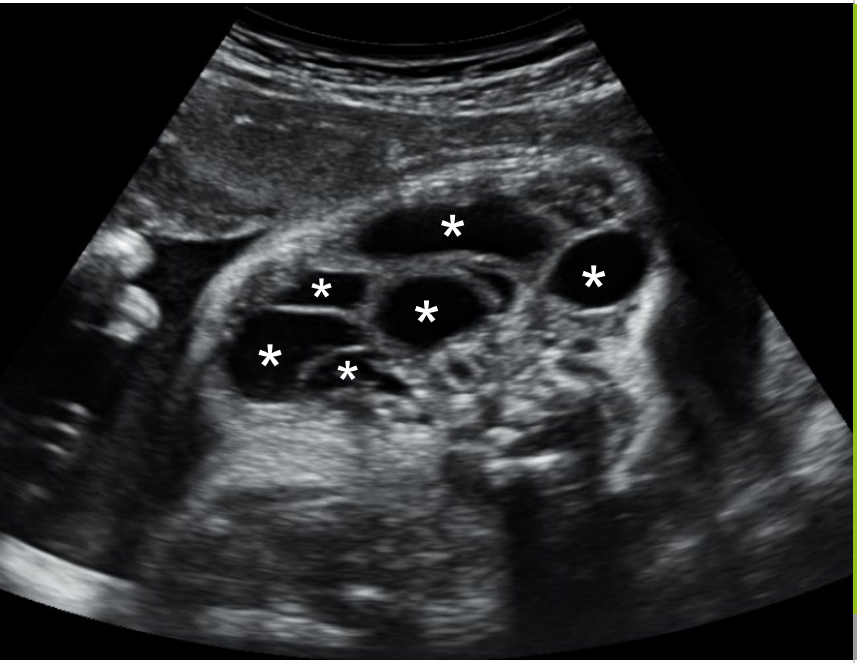


Fig. 2

Fetal ultrasound examination at 32 weeks of gestation: increasing size of the multicystic neck lesion (asterisks) and polyhydramnios (not shown) were documented.

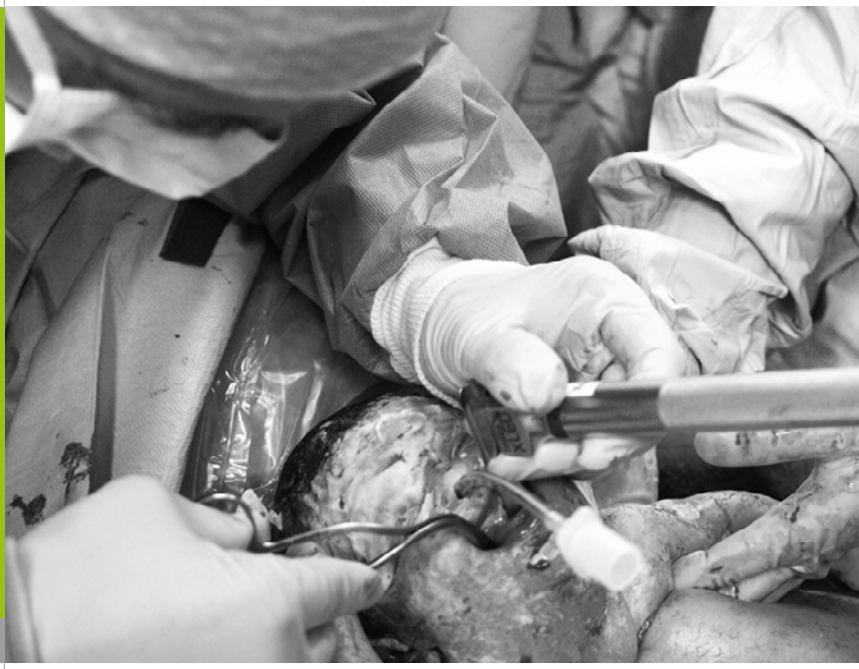


Fig. 3

EXIT procedure: successful nasotracheal intubation following partial delivery of the infant.

Fig. 4

EXIT procedure: following nasotracheal intubation, the endotracheal tube was sutured in place (A) and positive pressure ventilation was initiated with a self-inflating bag (B).

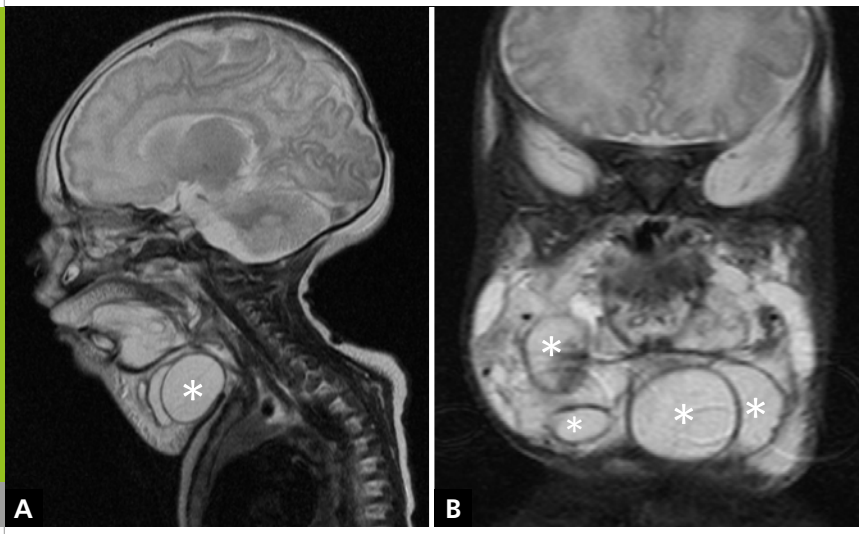


Fig. 5

Postnatal T2- weighted MRI demonstrating a large multicystic neck lesion (asterisks): A) sagittal view, B) coronal view.

The ex utero intrapartum treatment (EXIT) procedure was originally developed to reverse temporary tracheal occlusion in patients who had undergone fetal surgery for severe congenital diaphragmatic hernia (5). In recent years, it has increasingly been used to deliver babies who have airway compression due to cervical teratomas, cystic hygromas or blockage of the airway such as congenital high airway obstruction syndrome (CHAOS) (EXIT-to-Airway) (6). The procedure has also been used to stabilize and perform resection for babies with massive lung lesions (EXIT-to-Resection) and infants with severe forms of diaphragmatic hernia (EXIT-to-ECMO).

EXIT procedures are performed under maternal general anesthesia. A special uterine stapling device is used to prevent bleeding from the uterine incision, which could be excessive since the uterus is prevented from contracting. The baby is then partially delivered through the incision while remaining attached to the placenta. Once the airway is secured (in case of an EXIT-to-Airway procedure), oxytocin is administered and delivery is completed.

Most lymphatic malformations are benign lesions. In some cases, observation may be appropriate, as spontaneous regression has been reported to occur in up to 45% of cases. However, patients with lymphangioma colli frequently show signs of respiratory compromise and impaired swallowing; trauma (not seldom in active

toddlers), infection or intracystic hemorrhage are other potential complications that will lead to active interventions. Possible treatment options include sclerotherapy (4, 7) (Table), radiofrequency ablation, laser therapy or surgical resection. More recently, the use of propranolol (a well established therapy for infantile hemangioma), sildenafil and sirolimus has been reported (1). Unfortunately, lymphatic malformations have a high recurrence rate. In the future, better understanding of the genetics and biology of these lesions will hopefully lead to improved treatment strategies.

See also: COTM April 2006 «Intralesional injection therapy wit OK-432 (Picibanil®) in a full term infant with multicystic lymphangioma colli».

Agent	Assumed mechanism of action	Side effects / complications	Use in macrocystic LM	Use in microcystic/other LM
Picibanil® (OK-432), lyophilized mixture: Streptococcus pyogenes group A benzyl penicillin	Inflammatory response: increased cytokine production by leucocytes	Anaphylaxis	yes	no
Doxycycline, tetracycline antibiotic	Inhibition of matrix metalloproteinases and cell proliferation; suppression of VEGF-induced lymph-/angiogenesis; deposition of collagen and fibrin with cyst involution	Discoloration of teeth; electrolyte imbalances	yes	no
Bleomycin, chemotherapeutic agent	Inhibition of DNA synthesis; inflammatory reaction on endothelial cells	Interstitial pneumonia; pulmonary fibrosis (if cumulative IV dose > 400 mg)	yes	no
Pingyangmycin, chemotherapeutic agent	Destruction of lymphatic endothelial cells; increased collagen deposition in cyst cavity	Hair loss; gastrointestinal reaction; alteration of skin pigmentation; pulmonary fibrosis	yes	yes
Sodium tetradecyl sulfate (STS), detergent	Emulsion of cell membrane lipoproteins; increase of membrane permeability; enhancement of cell death and fibrosis, if used with doxycycline or ethanol	Increased risk of infection	yes	orbital
Ethanol, desiccant	Dehydration of lymphatic endothelial cells	Respiratory depression; cardiac arrhythmias; rhabomyolysis; hypoglycaemia; seizures	(yes)	no

Table. Sclerosing agents used in the treatment of lymphatic malformations (LM) (1).

REFERENCES

1. Defnet AM, Bagrodia N, Hernandez SL, Gwilliam N, Kandel JJ. Pediatric lymphatic malformations: evolving understanding and therapeutic options. Pediatr Surg Int 2016 Jan 27 (epub ahead of print) *(Abstract)*

2. Levy AD, Cantisani V, Miettinen M. Abdominal lymphangiomas: imaging features with pathologic correlation. Pictorial essay. AJR 2004;182:1485-1491 (no abstract available)

3. Lugo-Oliveieri CH, Taylor GA. CT differentiation of large abdominal lymphangioma from ascites. Pediatr Radiol 1993;23(2):129-130 *(Abstract)*

4. Rautio R, Keski-Kisula L, Laranne J, Laasonen E. Treatment of lymphangiomas with OK-432 (Picibanil). Cardiovasc Intervent Radiol 2003;26:31-36 *(Abstract)*

5. Harrison MR, Adzick NS, Flake AW, et al. Correction of congenital diaphragmatic hernia in utero VIII: Response of the hypoplastic lung to tracheal occlusion. J Pediatr Surg 1996;31:1339-1348 *(Abstract)*

6. Olutoye OO, Olutoye OA. EXIT procedure for fetal neck masses. Curr Opin Pediatr 2012;24:386-393 *(Abstract)*

7. Balakrishnan K, Menezes MD, Chen BS, Magit AE, Perkins JA. Primary surgery vs primary sclerotherapy for head and neck lymphatic malformations. JAMA Otolaryngol Head Neck Surg 2014;140:41-45 *(Abstract)*

concept & design by mesch.ch

SUPPORTED BY Vifor Pharma

CONTACT Swiss Society of Neonatology
www.neonet.ch
webmaster@neonet.ch