Pomeranian J Life Sci 2021;67(4):30-33 doi: 10.21164/pomjlifesci.761

Reconstruction of a large-sized defect in the upper abdominal wall with a porcine small intestinal submucosa biodegradable membrane in a newborn with Poland syndrome

Kaja Giżewska-Kacprzak[1, A] ✉, Krzysztof Mikulski[2], Hanna Chojnacka[1, B], Elżbieta Gawrych[1, C]

[1] Pomeranian Medical University in Szczecin, Department of Pediatric and Oncological Surgery, Urology and Hand Surgery, Unii Lubelskiej 1, 71-252 Szczecin, Poland
[2] Pomeranian Medical University in Szczecin, Department of Pediatric Intensive Care, Unii Lubelskiej 1, 71-252 Szczecin, Poland

[A] ORCID:0000-0002-5870-9464; [B] ORCID: 0000-0002-6567-8176; [C] ORCID: 0000-0002-6050-4046

✉ k.gizewska@gmail.com

ABSTRACT
Severe cases of gastroschisis require advanced and multi-staged surgical techniques to cover the abdominal wall defect enabling a secure covering of the eventrated organs. A biodegradable porcine small intestinal submucosa membrane is an attractive material that demonstrates the benefits of bioabsorbable materials. It has been used in children. There are only a few reports of severe gastroschisis treatment with this material. The aim of this work is to present a rare case of a giant upper abdominal wall defect in a newborn with Poland syndrome, treated surgically with a biodegradable membrane. The treatment protected the child from infection and enabled satisfactory healing. The use of the described material was adjusted to the severe general condition of the patient. The biodegradable porcine small intestinal submucosa membrane is a safe material to cover a large-sized abdominal defect in newborns.
Keywords: abdominal wall defects; porcine submucosa collagen graft; newborn; gastroschisis; biodegradable membrane.

INTRODUCTION

Children born with gastroschisis require immediate surgical intervention to secure the uncovered organs situated outside the abdominal cavity. Sometimes, severe stages of gastroschisis and other types of abdominal wall defects in newborns cannot be treated by primary closure. In such cases, eviscerated organs are placed into a silo bag – a plastic tube that covers the organs and is hung over the newborn. In many cases the natural pressure of gravity allows the walls of the abdominal cavity to adjust its reduced capacity [1]. However, if secondary closure is still impossible after approx. a week, there are several options available. Prosthetic patches may cover the defect but don't allow definitive closure and will eventually need to be removed. A biodegradable porcine small intestinal submucosa (SIS) membrane is an attractive material that demonstrates the benefits of bioabsorbable material for example, in bladder augmentation [2]. It has been used in children. There are only a few reports of severe gastroschisis treatment with this kind of material [3, 4]. We have not found any publications about defects in the upper part of the abdominal wall. Moreover, there have been no publications about this topic in Polish journals.

The aim of the work is to present a rare case of a giant abdominal wall defect in a newborn treated surgically with a porcine intestinal submucosa biodegradable membrane. The use of the bioabsorbable material was adjusted to account for the severe condition of the patient who additionally presented symptoms of Poland's syndrome.

CASE REPORT

A 31-week gestational age female infant (body mass 2,800 g, Agar 2, 3, 4) had a giant upper abdominal wall defect exposing the whole liver and part of the large and small bowel (Fig. 1). Moreover, the baby presented asymmetry of the breast (the right side was significantly more underdeveloped than the left), lack of a right nipple and mild contraction of the fingers of the right hand. Severe respiratory failure required intubation immediately after birth. Due to the large size of the abdominal wall defect and small abdominal cavity with bowel and liver evisceration, primary closure was impossible. As the patient required initial intensive care stabilisation, the surgery was postponed to the 2nd day of life. The eviscerated organs were placed in a „hand-fashioned Silo Bag'' (Fig. 2). On day 18, the pouch was removed and a Gore-Tex patch was sewn into the edges of the defect as closure was impossible. Based on the poor stretching potential of the remaining abdominal wall and the risk of synthetic prosthesis infection, a biodegradable porcine submucosa membrane was chosen for the definitive closure of the defect. On the 36th day of life, the Gore-Tex patch was removed and a biodegradable patch made of SIS – Biodesign 4-Layer Tissue Graft was sewn in (Fig. 3). The membrane was covered with moistened surgical dressings. A state of chronic pneumonia, multiple saturation drops and bradycardia, severe obstructive and atelectatic-inflammatory changes persisted despite intensive and long-term treatment. Three sudden cardiac arrests in intervals of several days were managed by direct and pharmacological resuscitation. The

FIGURE 1. A large-sized upper abdominal wall defect presented after birth exposing the liver, stomach and the majority of the bowel loops

FIGURE 2. A hand-fashioned Silo Bag implemented in the first hours of the newborn's life

treatment included combined antibiotic therapy (repeatedly modified), immunotherapy, prophylaxis of bleeding, diuretics, catecholamines, and correction of water and electrolyte disturbances. During hospitalization in the Neonatal Intensive Care Unit, complementary blood transfusions were performed 7 times due to persistent anaemia. Total parenteral nutrition was used until the 68th day of life.

FIGURE 3. A porcine small intestinal submucosa biodegradable membrane covering the defect

At 4 months of age, the infant was transferred to the Intensive Care Unit for Children to continue the therapy. An X-ray and computed tomography (CT) of the chest showed changes typical of bronchopulmonary dysplasia and an abnormal bone structure of the chest (Fig. 4). The ribs on the right side were distorted and dislocated, the right scapula was shorter and deformed in the area of the lower edge, the xiphoid process of the sternum was deformed and the greater pectoral muscles on the right side were hypoplastic. Moreover, a mild contracture of the fingers of the right hand was observed. The CT showed a large defect of 10 cm in the abdominal wall, located on the right side. The right lobe of the liver was placed on a III to V rib outside of the abdominal cavity. The small intestine and cecum were also displaced outside. Additionally, the inferior vena cava and an additional renal artery were displaced to the right and described as stenotic. Echocardiography revealed an atrial septal aneurysm with PFO/ASD II.

Due to the concomitant abnormalities of the skeletal system suggesting the coexistence of Poland syndrome, genetic counselling was recommended. A deletion in the 2q37.3 region was confirmed, which, according to the International Standards for Cytogenomic Arrays, may have a mild effect on the patient's phenotype and may be correlated with developmental delays. It was suggested that additional developmental disorders conditioned by isolated external mechanical or environmental factors or in combination with genetic factors may have played a key role in the formation of Poland syndrome diagnosed in this child.

Within 3 months after implementation of the biodegradable patch, the natural material was completely healed and covered with the patients' own epidermis (Fig. 5). There was an obvious defect in the musculature of the abdominal wall that created a wide hernia. Nevertheless, there has been no gastrointestinal complications up to date. The child tolerated oral feeding well.

As prolonged mechanical ventilation was anticipated, a tracheostomy was created. Repeated attempts to get the patient off the

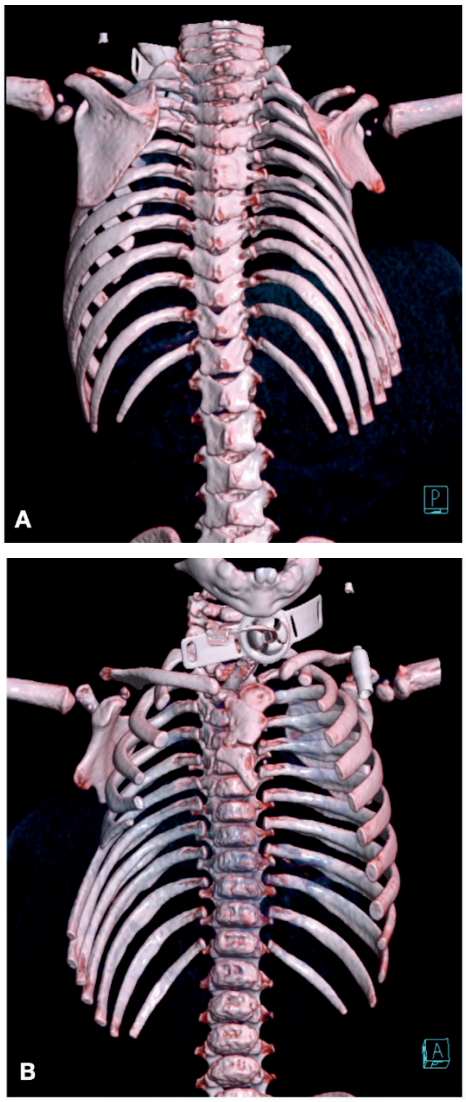

FIGURE 4. Chest computed tomography 3-dimensional reconstruction: A) right scapula and rib deformity; B) sternum deformity with decreased right chest cavity volume

FIGURE 5. Long term follow-up in the second year of the patient's life

ventilator failed. A worsening of the respiratory function and fatigue was observed. After 18 months of continuous ventilation, an attempt was made to switch on alternating ventilation from a „home" ventilator, alternating with self-breathing, with good results. Neurological rehabilitation and learning to walk in a walker were introduced.

After 20 months of hospitalization, the child was discharged home to the care of her parents. The tracheostomy was closed 3 months after the initial discharge.

In the future, plastic surgery of the abdominal wall should be very carefully considered because the reduction of the abdominal cavities capacity with a smaller capacity of the right side of the chest may result in symptoms of reduced respiratory function in the patient.

DISCUSSION

The optimal features of a biodegradable implant include good healing potential with possible minor tissue reaction, resistance to infection, its ability to permanently connect to the patient's tissue, durability and flexibility of its structure and a lack of adhesive reaction. From a surgical point of view, one of the most important features is the smooth internal surface of the material structure adjacent to the organs of the peritoneal cavity, which significantly reduces or eliminates the risk of adhesion to internal organs [5].

The research on prosthetic material for the repair of abdominal defects has evolved over the past several years. The ultimate goal is a biomaterial that heals spontaneously without adverse sequelae. Synthetic materials, although available, seem to be inferior to biological polymers because of the significant adhesion formation and the fact that synthetic material does not grow with the infant [6]. The interest in biological matrices is growing as there are limitations to synthetic meshes. Biological scaffolds are materials composed of extracellular matrices derived from a variety of tissues of human or other mammalian origin [7, 8].

One of the directions in the search for a perfect material for covering defects are experimental animal models. Drewa et al. used tissue engineering to repair the abdominal wall in an animal model. The defect was covered with a polyglycolic acid scaffold with seeded fibroblasts. Histological and general examination revealed good neovascularization in the tissue walls of the abdominal cavity and no intense scars between the abdominal wall and the skin. The authors suggest that the angiogenesis process is sufficient to obtain the tensile strength and plasticity of the graft and is one of the main factors responsible for their later part [8].

A porcine SIS biodegradable membrane has been rarely reported in newborns. Karpelovsky et al. presented a series of children that required the biodegradable material to support the definitive closure of the abdominal wall after liver or kidney transplants [4]. When looking for the best solutions for our patient, we found a case series by Gabriel and Gollin in which 3 newborns with complicated gastroschisis required a porcine SIS biodegradable membrane [3]. The authors presented very satisfying results. Their protocol included a negative pressure

dressing and further resection of the "neofascia" resulting in a durable abdominal wall reconstruction and umbilical hernia correction. In the present case, the defect was extremely large and located in the upper part of the abdomen affecting the right side of the lower chest as well. The unusual clinical features of the underdevelopment of the right upper part of the thorax and upper limb draw suspicion to the presence of Poland syndrome – a rare syndrome with an estimated occurrence of 1:10,000 to 1:100,000 [9]. A final diagnosis could have been reached with further genetic tests but the parents did not continue to follow diagnostic measures. Our patient was disqualified from the negative pressure dressing as she was generally unstable and because both the liver and right lung could have been affected. We believe that the long-term healing effect resulting in the epithelization of the membrane, or "neofascia", was the best possible result while at the same time the least invasive for our patient. We are happy to present our case as an example of how new surgical material can be implemented in the youngest strata of the population with the required adjustments.

CONCLUSION

The use of a porcine SIS biodegradable membrane in the technique of closing a large abdominal defect in a newborn is a safe approach. It can be adjusted to the severity and localization of the defect.

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