



## A Review on Pectus Excavatum in Canines: A Congenital Anomaly

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### Abstract

Pectus Excavatum is a congenital developmental deformity of the anterior chest wall, characterized by the dorsal deviation of the caudal sternum and associated costal cartilages or a ventral to dorsal narrowing of the entire thorax in which several ribs and the sternum grow abnormally. It has been reported in dogs, kittens, lambs and calves. Burmese kittens and Brachycephalic dogs are more predisposed. Common clinical signs include increased inspiratory effort, inspiratory stridor, moist rales, dyspnea and exercise intolerance. Cardiac murmurs associated with concurrent cardiac defects or compression of the heart and kinking of the great vessels are common. Increased pressure in utero, rickets and increased traction on the sternum due to abnormalities of the diaphragm have been postulated as specific mechanisms. Pectus Excavatum is initially suspected from visual examination of the anterior chest. The radiographic confirmation of Pectus Excavatum is based on the thoracic shape and radiographic changes. Cardiac malposition is usually seen, with the heart shifted to the left of midline and sometimes cranially. More objective parameters have also been suggested including the Fronto-sagittal index (FSI) and vertebral index. More recently the "Haller index" has been used based on CT scan measurements. An index over 3.25 is often defined as severe. Patients with mild disease (flat chest) may become normal without surgical intervention. However, animals with moderate or severe disease need surgical intervention. Severity of this condition is repaired in animal by surgical removal of the affected portion of the sternum and replacement with a graft and also by using a cast with sutures around the sternum.

**Key words-** Pectus Excavatum, Canine, Congenital Anomaly

### Introduction

Pectus Excavatum (Latin meaning Hollowed Chest) is the most frequently observed congenital deformity of the anterior chest wall, characterized by the dorsal deviation of the caudal sternum and associated costal cartilages or a ventral to dorsal narrowing of the entire thorax in which several ribs and the sternum grow abnormally<sup>1,2</sup> producing a caved-in or sunken appearance of the chest and has been reported in both dogs and cats.<sup>1,3,4</sup> Pectus Excavatum is sometimes referred to as Cobbler's chest, Sunken chest, Funnel chest or simply a Dent in the chest.<sup>5</sup> Among the thoracic wall deformities resulting in a change in rib shape, Pectus Excavatum is the only one described with any frequency in the veterinary literature.<sup>6</sup> Pectus Excavatum has also been reported in dogs<sup>7</sup>, lambs

and calves<sup>8</sup> and sea otters<sup>9</sup>. Although occurring less commonly than in man, sixteen cases were reported in cats<sup>10-14</sup>, nine were in domestic short hairs (DSH), four in Orientals, two in long hairs and one in a British shorthair, of which DSH group comprising 7 males and only 2 females.<sup>1</sup>

The costo-sternal deformities resulting in dorsoventral narrowing of the chest, primarily in the caudal aspect, may have secondary abnormalities of respiratory and cardiovascular function from restriction of ventilation and cardiac compression.<sup>15</sup> Mostly the cases are congenital however it may develop at puberty also. Acquired disease is seen in humans when large, negative intrapleural pressures on compliant chest walls result in the collapse of the sternum and intercostal cartilages. Speculation exists in veterinary medicine that upper respiratory obstruction at a young age may cause abnormal respiratory gradients and subsequent Pectus Excavatum. Some patients demonstrate the "swimmer syndrome" in which neonatal dogs lack the ability to posture properly and remain in sternal recumbency leading to invagination of the sternum. Patients with such abnormality

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may experience negative psychosocial effects and avoid activities that expose the chest.

### Abnormalities Affecting the Rib Cage and Sternum

A variety of rib deformities are encountered and are not uncommon, including missing ribs (usually T13), fused ribs, extra ribs (usually L1) and malformed ribs. Pectus Carinatum (chicken breast) is a congenital abnormality that results in a laterally compressed thorax secondary to ventral displacement of the caudal aspect of the sternum; reported in animals. The two most common congenital defects are flat-chested kitten syndrome (FCKS) and Pectus Excavatum (PE). PE affects the sternum and costal cartilages resulting in ventro-dorsal narrowing of the thorax whereas FCKS affects the whole rib cage resulting in a dorso-ventral flattening of the thoracic cavity.

### Signs and Symptoms

- Respiratory problems are common (increased inspiratory effort, inspiratory stridor and moist rales) are common, if infection persists. Respiratory distress results from restricted ventilation and paradoxical movements of the deformity during inspiration. Chronic alveolar collapse contributes to exercise intolerance and hypoxia by impairing gas exchange.
- Dyspnea, exercise intolerance, weight loss, hyperpnoea, recurrent pulmonary infections, cough, vomiting, cyanosis, poor appetite and episodes of mild upper respiratory disease.<sup>2,16</sup>
- Heart sounds are often muffled, especially over the right hemithorax. Cardiac murmurs associated with concurrent cardiac defects or compression of the heart and kinking of the great vessels are common. Cardiac displacement and apparent cardiomegaly occurs in animals.<sup>4</sup>
- There is no correlation between severity of clinical signs and the severity of anatomic or physiological abnormalities. The thoracic defect is easily palpated or seen on physical examination.<sup>17</sup>

### Cause and Risk Factors

- A genetic predisposition may exist. Approximately 37% of individuals with Pectus Excavatum have a first degree family member with the condition. Burmese kittens and Brachycephalic dogs are more predisposed, suggesting a possible heritable basis for defect in small animals.<sup>18</sup>
- Dogs predisposed to respiratory obstructive processes, suggesting the elevated pleural pressure from chronic elevation of upper airway resistance may contribute to the development of the condition
- Physiologically, increased pressure in utero, rickets and increased traction on the sternum due to abnormalities of the diaphragm have been postulated as specific mechanisms.<sup>19</sup> Patients with Spinal muscular atrophy, defective Osteogenesis and

Chondrogenesis or a combination of these has also been postulated.<sup>20</sup>

- Puppies raised on surfaces with poor footing may be predisposed to "swimmer's syndrome." Most commonly presented between 4 weeks and 3 months of age.

### Aetiology

The aetiology of the condition is unclear and may well involve multiple causes. Theories including abnormal pressure gradients in brachycephalic dogs or in upper respiratory tract obstruction, shortening of central tendon of diaphragm, abnormal intrauterine pressure, deficient muscular components derived from the septum transversum of the diaphragm, congenital thickening of the musculature of the cranial portion of the diaphragm, thickening of substernal ligament, failure of osteogenesis/chondrogenesis, arrested sternal development and rachitic influences were reviewed.<sup>12</sup> Pectus Excavatum has also been reported associated with mucopolysaccharidosis in a cat.<sup>21</sup> Pectus Excavatum can occur as an autosomal dominant condition in man and has been reported in litter of setter cross puppies.<sup>17</sup>

### Diagnosis

Pectus Excavatum is initially suspected from visual examination of the anterior chest. Auscultation of the chest can reveal displaced heart beat and valve prolapse. There can be a heart murmur occurring during systole caused by proximity between the sternum and the pulmonary artery. Lung sounds are usually clear yet diminished due to decreased base lung capacity.<sup>22</sup> The diagnosis of PE is based on the thoracic shape and radiographic changes. More objective parameters have also been suggested including the fronto-sagittal index (FSI) and vertebral index. Many scales have been developed to determine the degree of deformity in the chest wall. Most of these are variants on the distance between the sternum and the spine. One such index is the "Backer ratio" which grades severity of deformity based on the ratio between the diameter of the vertebral body nearest to xiphosternal junction and the distance between the xiphosternal junction and the nearest vertebral body.<sup>23</sup> More recently the "Haller index" has been used based on CT scan measurements. An index over 3.25 is often defined as severe.<sup>24</sup> The Haller index is the ratio between the horizontal distance of the inside of the ribcage and the shortest distance between the vertebrae and sternum.<sup>25</sup> Some studies also suggest that the Haller index can be calculated based on chest x-ray as opposed to CT scanning in individuals who have no limitation in their function.<sup>26</sup> Chest x-rays are also useful in the diagnosis.

The chest x-ray in Pectus Excavatum can show opacity in the right lung area that can be mistaken for an infiltrate.<sup>27</sup> Radiographs confirm the diagnosis. Deformities that cause a decreased thoracic volume are readily noted. Cardiac malposition usually is seen, with the heart shifted to the left of midline and sometimes cranially. Cardiac enlargement may be detected. Lung fields may show evidence of concurrent

disease. Shifting of the heart shadow to the left may expose the right hilus and encourage a diagnosis of pulmonary disease. Echocardiography is indicated to fully evaluate cardiac status (eliminate primary cardiac disease and detect possible concurrent cardiac defects). Cardiac enlargement on radiographs may be attributed only to malpositioning.

### Differential Diagnosis

Pectus Excavatum is differentiated from other disorders by a series of elimination of signs and symptoms. Pectus Carinatum is excluded by the simple observation of a collapsing of the sternum rather than a protrusion, also known as Pectus Galinatum or pigeon breast.<sup>28</sup> Kyphoscoliosis is excluded by diagnostic imaging of the spine as the spine in Pectus Excavatum is usually normal in structure necessitating the importance to differentiate Pectus Excavatum from paradoxical movements of the sternum in a young animal with respiratory distress and profound respiratory effort.<sup>4</sup> Numerous causes of dyspnea, cyanosis, hyperpnoea, and/or cough must be entertained. Physical examination and radiographic evaluation enables the clinician to rule out the most common differentials:

- Tracheal malformations or collapse
- Cardiac disease
- Electric cord bite
- Hemothorax
- Pyothorax
- Pneumonia
- Allergic bronchitis
- Stenotic nares
- Elongated soft palate

### Treatment

The decision for surgical repair of deformities should be made on the basis of clinical signs of disease. Patients with mild disease (flat chest) may become normal without surgical intervention. Manual medial compression of the thorax by the owners or in the form of a splint is recommended. Animals with moderate or severe disease are surgical candidates. Fronto-sagittal and vertebral indices provide objective criteria for determining severity. The technique for repair may be dictated by the age of the animal (young animals with a compliant sternum and ribs may do well with external coaptation; older animals with less compliant thoraces may need partial sternectomy).

Pectus Excavatum-associated cardiopulmonary dysfunction may be life threatening. Several surgical techniques including both closed and open methods of correction have been described in both dogs and cats. The use of U-shaped external splint (X-Lite classic splint; EBI Biomet, USA) has been previously reported.<sup>4,14</sup>

Before an operation proceeds several tests are usually to be performed. These include, but are not limited to, a CT scan, pulmonary function tests, and cardiology exams (such as auscultation and ECGs).<sup>15</sup> After the CT scan is taken the

Haller index is measured. A Haller Index of greater than 3.25 is generally considered severe; while normal chest has an index of 2.5.<sup>25</sup> The cardiopulmonary tests are used to determine the lung capacity and to check for heart murmurs.

The fronto-sagittal and vertebral indices are important in defining the degree of the deformity (mild, moderate or severe), as well as the response to the treatment.<sup>2,4</sup> Based on suggested values for determining the severity of the Pectus anomaly<sup>4</sup> as well as on the intensity of clinical signs, characterizes as moderate (frontal index: 2.0–3.0; vertebral index: 6.0–9.0) and mild as (frontal index: < 2.0; vertebral index: > 9). Normal non brachycephalic dogs have a fronto-sagittal index of 0.8 to 1.4 and a vertebral index of 11.8 to 19.6.

### Surgical Technique (Canines)

Some procedures used to treat the condition in humans have not been used in animals, such as the use of a cast with sutures wrapped around the sternum and the use of internal and external splints.<sup>13,29</sup> These techniques are generally used in immature animals with flexible cartilage. There are two ways; this condition is repaired in animals:

Surgical removal of the affected portion of the sternum and replacement with a graft.

2. Use of a cast with sutures around the sternum.

Solid customized and prefabricated silicone implants have been used in wide range of congenital or acquired chest wall deformities seeking to augment a muscularly deficient or underdeveloped chest. The immediate postoperative problem of seroma and subcutaneous implant show has been minimized by careful planning, gentle technique, deep insertion, improved patient positioning on the operating table, and the use of oral anti-inflammatory medications. The long-term results of these implants seem very satisfactory. The patients are usually physically active, and the implants show no long-term sequelae such as seroma, infection, displacement or rupture.<sup>30</sup> Three types of surgical repair for Pectus Excavatum have been described in dogs and cats.

1. External Splinting

2. Internal Splinting

3. Longitudinal Sternebral Pining combined with External Splinting.

In the external splinting type, moldable splinting material is used to contour a U-shaped or V-shaped splint.<sup>4,13,14</sup> In the internal splinting type, veterinary cuttable plate or aluminum splint rod is used to realign a noncompliant sternum. This technique requires the exposure of the site of sternal deviation and placement of the plate after correction of the deformity.<sup>16,29</sup> In the longitudinal sternebraal pinning combined with external splinting, moldable splinting material and a Kirschner pin are used to realign an on compliant sternum. In this technique, a Kirschner pin is inserted through the sternum from the manubrium to fourth sternebra.<sup>14</sup>

Some anesthetic precautions are necessary during the surgical procedure for treating Pectus Excavatum due to the age of the animal and the respiratory distress, such as avoiding an extended period of fast, maintaining body

temperature, using anesthetic drugs that are easily metabolized, and using endotracheal intubation to provide airway access.<sup>24</sup>

First a fiberglass cast is molded to a normal shape of the bottom of the chest. Then sutures are placed around the affected sternabrae, start caudally and the sutures are placed; pull up on the preceding suture so that it will pull the sternum away from the lungs and heart, as the next suture is placed, a piece of plastic (plastic bag) that has the same dimensions as the cast is placed exactly in position on the chest as the cast will normally be seated (the two black lines on the chest denote the hind extend of the cast); markings are made on the plastic template indicating where the sutures should run through the cast.

The use of U-shaped external splint (X-Lite classic splint; EBI Biomet, USA) has been previously reported. The successful surgical correction of Pectus Excavatum using U-shaped external splint and cylindrical external splint (Orthoplast; Johnson & Johnson, USA) in the cats has been described.<sup>31</sup> In this study prior to surgery, a cylindrical external splint for and U-shaped external splint were contoured to the normal shape of the patient's thorax. The patients were positioned in dorsal recumbence. Stay sutures were placed around the sternabrae using 0-No polypropylene (Ethicon, USA) from the manubrium cranially, to the xiphoid caudally. The suture ends were left long and tagged with mosquito hemostats. All stay sutures were passed through the holes on the apex of the splint using an 18-gauge needle and then tied securely. The splint was held in place with the umbilical tape at the cranial aspect, acting as shoulder straps. Velcro straps were placed dorsally to ensure proper splint positioning.

Corrective treatment using an external splint may alleviate the impaired ventilatory performance when done in young animals, since the costal cartilages and sternum are pliable.<sup>1,13</sup> Possible complications with the method are damage to the internal thoracic vessels, heart, or lungs during the passage of the needle around the sternum; fatal reexpansion due to pulmonary edema<sup>16</sup> and postoperative skin abrasions, suture abscesses, and dermatitis. A pin inserted longitudinally through the manubrium in association with an external splint<sup>14</sup> or a plate applied to the ventral side of the sternum<sup>29</sup> are alternative techniques that have been reported for use in young cats. In the case of older animals or those treated unsuccessfully with external splinting, surgical procedures, such as partial sternectomy or ostectomy with external fixation, may be used.<sup>1</sup>

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## چکیده

### مروری بر Pectus Excavatum در سگ: یک آنومالی مادرزادی

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بخش جراحی و رادیولوژی دانشکده دامپزشکی، دانشگاه علوم کشاورزی و تکنولوژی کشمیر، کشمیر، هند

Pectus Excavatum یک دفرمیتی مادرزادی تکاملی دیواره قدامی قفسه سینه است که با انحراف پشتی قسمت خلفی جناغ و غضروف-های دنده‌ای مربوطه یا تنگی شکمی تا پشتی کل ناحیه توراکس مشخص می‌شود بطوریکه چندین دنده و استخوان جناغ بصورت غیرطبیعی رشد کرده‌اند. این عارضه در سگ، گربه، بره و گاو گزارش شده است. گربه‌های نژاد Burmese و سگ‌های براکی سفالیک بیشتر مستعد هستند. نشانه‌های بالینی معمول شامل تلاش در تنفس دمی، سختی تنفسی، رال تنفسی و عدم تحمل به ورزش است. مرمز قلبی مرتبط با نقایص قلبی یا بدلیل فشردگی قلب و انحنای عروق بزرگ معمول است. افزایش فشار در دوران جنینی، ریکتز و افزایش کشیدگی جناغ بدلیل مشکلات دیافراگم بعنوان مکانیسم‌های اختصاص مطرح است. این دفرمیتی در ابتدا با معاینه بصری قسمت قدامی قفسه سینه مورد شک قرار می‌گیرد. تایید رادیوگرافی این عارضه با مشاهده تغییرات رادیوگرافی و تغییر شکل قفسه سینه می‌باشد. معمولاً قلب در جای طبیعی خود قرار ندارد و به سمت چپ قفسه سینه و گاهی به سمت قدام جابجا می‌شود. پارامترهای پیشنهاد شده شامل اندیس (FSI) Fronto-Sagittal و اندیس مهره‌های (Vetebral Index) می‌باشد. اخیراً اندیس هالر (Haller Index) براساس اندازه‌گیری‌های CT-Scan استفاده می‌شود. اندیس بالاتر از ۳/۲۵ اغلب نشانه شدید بودن عارضه است. بیماران با بیماری ملایم (قفسه سینه صاف) ممکن است بدون عمل جراحی نرمال باشند. اگرچه حیوانات با بیماری متوسط و شدید نیاز به دخالت جراحی دارند. شدت این عارضه در حیوانات با برداشتن قسمت درگیر جناغ و جایگزینی با یک پیوند و همچنین با استفاده از یک قالب با بخیه زدن آن به اطراف جناغ اصلاح می‌شود.

کلمات کلیدی - Pectus Excavatum، سگ، آنومالی مادرزادی