Congenital Hyperrotated Colon

Francisco T. Tirol, MD, FASAS

Pioneers Memorial Healthcare District
Brawley, CA

Address for correspondence:
Francisco T. Tirol, M.D.
PO Box 439
El Centro, CA 92244-0439
Tel: (760) 352-6088
Fax: (760) 353-1533
Email: fttirol@aol.com

ABSTRACT
The congenitally hyperrotated cecocolon is a fact that should be established as a diagnosis. It is one of the conditions that lead to recurrent cecocolic torsion. Being preternaturally mobile it has the propensity for torsion. It is estimated to occur in eleven to twenty per cent of the population without gender preponderance. The common presentation is a subacute and chronically recurrent torsion and obstruction with spontaneous resolution. It may proceed into an acutely obstructing and strangulating event. It is often a missed diagnosis. Many of its baffling symptomatology is shared in common with other variants of recurrent cecocolic torsion but with the awareness to this diagnosis, its nuanced symptomatology is easily defined. An inductive review of the pathology, the pathogenesis, and a photographic documentation as the basis for its diagnosis is discussed.

INTRODUCTION
The congenital hyperrotated colon is not as rare nor as benign as we are led to believe.[1,2,3] This elongation is a developmental over-rotation which occurs in-utero during the second and third stage of colon differentiation.[4] The elongation results in the absence of fusion to the parietal wall. It imbues the cecocolon with preternatural mobility and predisposes it to torsion and obstruction.

The acute obstructive form manifests with symptoms of severe expectation and unequivocal surgical indication. The chronic and recurrent obstructive symptom complex can be nuanced and baffling to define. The presentation may be similar to those of the normally rotated but mobile cecocolic variants. The diagnosis can be elusive and challenging to define. Awareness to the diagnosis is at a premium.

An inductive approach by reviewing the embryology, pathology, and the pathogenesis as the basis for the nomenclature, diagnosis, and treatment is discussed.

DISCUSSION
A right lower quadrant abdominal pain caused by a symptomatic hyperrotated cecocolon is usually never a primary nor even a differential diagnostic consideration.

This particular entity is often misdiagnosed by those who have been trained to habitually and empirically call it as a cecal volvulus.[1,2,6,7] It would be of no consequence if the torsion and obstruction of the cecocolon is of only one type but this is not so because there are many variants under this heading. This is an unfortunate insularity which continues to be perpetuated to this day, not only by the clinician but also by the radiologist.[1,2,7]

The completion into a three hundred sixty degrees of torsion by the cecum is an anatomical impossibility. Unlike the cecal bascule, a cecal volvulus is therefore an anatomical misnomer. Furthermore, It is a nominal generalization which obligatorily encompasses other variants of cecocolic abnormalities which are specific diagnoses per se.[5] This diagnostic misidentification often leads to the unintended consequence manifested by undue suffering of the undiagnosed chronically symptomatic patient. These patients are often alleviated only after the acute obstructing and strangulating episode has exposed the indisputable symptoms of an irreversible obstruction and strangulation leading to a mandatory surgical correction.

For obscure reasons, the radiologist who often sees a hyperrotated colon does not report. They avowedly see this anomaly routinely but perhaps they do not attribute any clinico-pathological correlation to it. It bears mention that there is also the normally rotated but mobile cecocolon. These morphological variants have gained clinical recognition because they are frequently involved in recurrent cecocolic torsions with spontaneous resolutions,[4,5] and they may manifest the same symptoms as the hyperrotated variant.

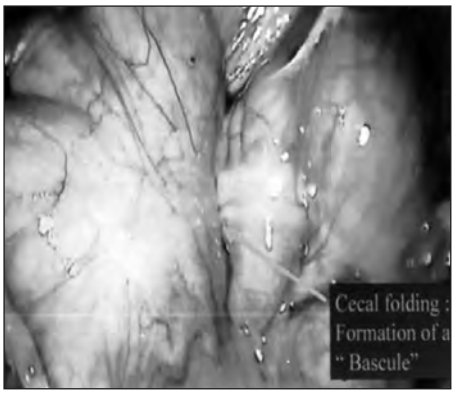

FIGURE 1. Cecal Bascule

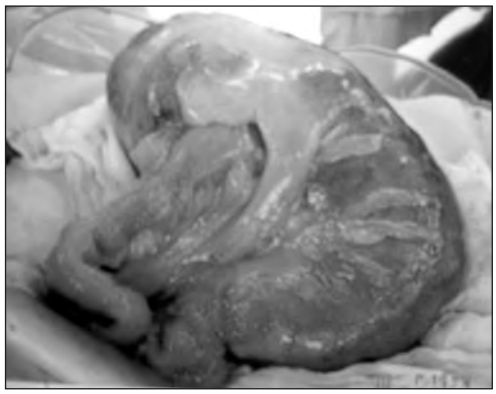

FIGURE 2. Acute Cecocolic Torsion. Note untorsed ileum

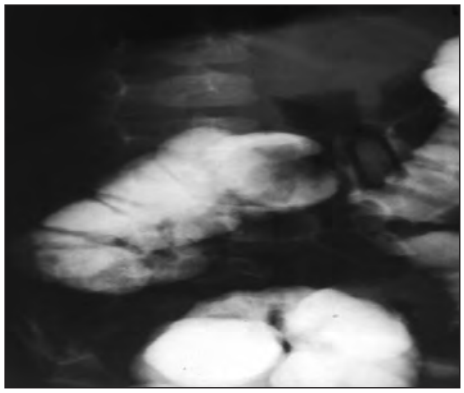

FIGURE 3. Hyperrotated Cecocolon in 16 month old child with currant jelly stools, detorsed with gastrografin enema

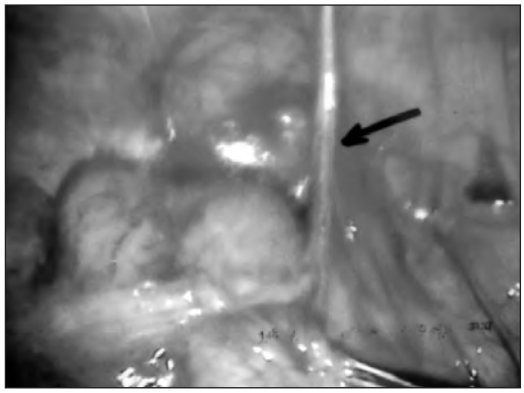

FIGURE 4. Funicular Jackson's Membrane

NOMENCLATURE

A normally rotated but seesawing mobile cecum is called a cecal bascule[8] (Figure 1). When a normally rotated or hyperrotated cecum and ascending colon is involved in recurrent torsion it has been called a floppy cecum syndrome.[9] Any of the foregoing variant that results in obstruction-strangulation has been called a cecal volvulus. This terminology, which is an anatomical misnomer, is still fashionable and the concept continues to be restated by more recent workers.[7]

A preternaturally mobile cecocolon may fold, twist and torse incompletely. Anatomically it can torse completely three hundred sixty degrees into a volvulus only if it incorporates the terminal ileum with its mesentery. In a midgut volvulus can this condition may occur. Should the torsion of this segment completes into a volvulus, it can only be an ileocecocolic volvulus. Insofar as the author is aware, there is no specific genre of this type of volvulus that has yet been documented visually even up to this era of copious photographic availability.

Similar to the cecocolic torsion presented (Figure 2), close scrutiny of a photograph in a surgical text of a purported cecal volvulus actually depicts an acute cecocolic torsion.[10] The term floppy cecum syndrome is best avoided because it does not signify any pathological significance and anatomically it mentions only the cecum and omits mentioning the ascending colon.

INCIDENCE

The incidence of the congenitally hyperrotated colon has not been verified at autopsy unlike the normally rotated mobile cecocolon which was documented to occur in about ten to twenty per cent of the population.[6] There was no mention whether these cases included the hyperrotated variant nor was gender preponderance mentioned. This rate has been extrapolated to be one in twenty to twenty four thousand of the population. It is twenty times less common than acute appendicitis. About twenty percent of these cases is believed to be symptomatic.[11] It is fair to believe that this anomaly can occur at any age. A sixteen month child with recurrent colic for months eventually presented doubled-up in severe colic with currant jelly stools. He was successfully detorsed with water soluble contrast[5] (Figure 3).

EMBRYOLOGY

The first stage of the colonic differentiation and development of the midgut is extra-abdominal and starts from the sixth to tenth week "in utero". The second stage is the continuing development of the midgut with colon differentiation and rotation two hundred seventy (270) degrees while being drawn intra-abdominally until approximately the eighth month of pregnancy. Rotation terminates after the cecum locates at the right iliac fossa. The third stage is cecocolic fixation to the parietal peritoneal wall at the right paracolic gutter by a mesocecocolon which ends at about the fourth month post partum.[12]

There may be a Jackson's Membrane at the cecum[13] (Figure 4) or a Parietocolic Band or Membrane[14] (Figure 5) at the distal third of the ascending colon. A Lane's band may be present at the distal ileum.[13] These membranes range from being filmy and velamentous to funicular. They have been mistaken for adhesions.[13] Cecal folds which tethers the cecum to the parietal wall may be present.[15]

The midgut is supplied by the superior mesenteric artery which gives rise to the ileocolic and right colic arteries. These supply the ileum, cecum, appendix, and ascending colon. Specifically, the cecum has a double blood supply consisting of the anterior and posterior cecal arteries. They are not end arteries and are anastomotic to each other in uninterrupted sequence aided by the marginal arteries of Drummond. The innervation comes from the coeliac ganglion.

PATHOLOGY

An extra-abdominal maldevelopment results in an Omphalocoele.[12] A rotational arrest during the second stage results in underrotation of the colon, customarily called malrotation. There may be an associated Ladd's band which can cause a high intestinal obstruction by constricting the descending or second segment of the duodenum.[16,17] A normally rotated cecocolon may have an arrested third stage by nonfixation of the cecum or the

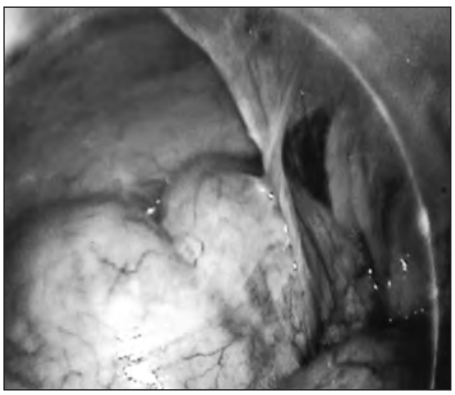

FIGURE 5. Funicular Parietocolic Membrane

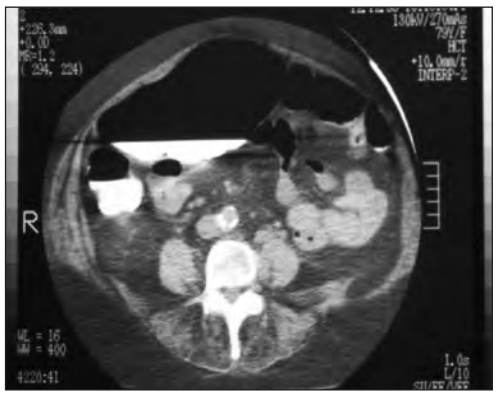

FIGURE 6. Cecocolic Pseudotumor by CT scan. Normal ascending colon and small intestine (by barium enema).

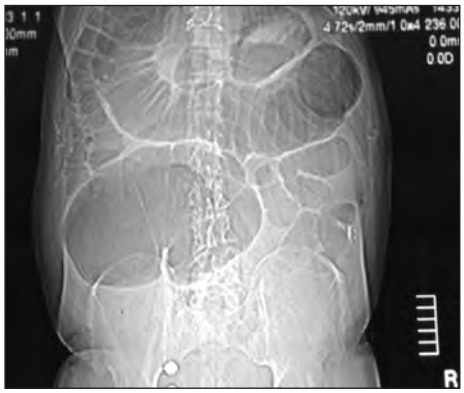

FIGURE 7. Cecocolic Pseudo-obstruction: CT scan: Note gas distal to site of obstruction.

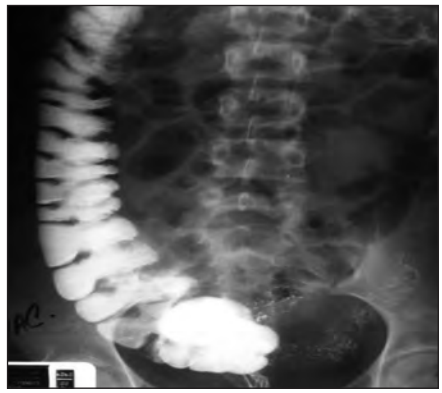

FIGURE 8. Hyperrotated Cecocolon in true pelvis: with dyspareunia

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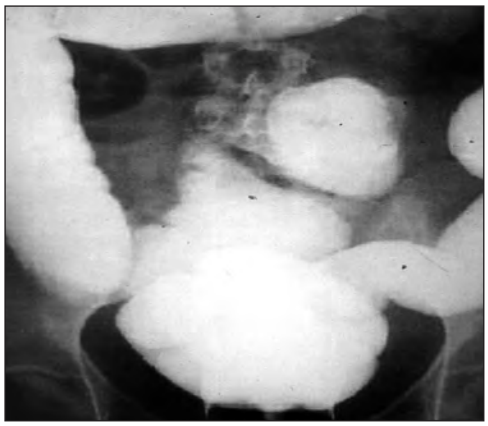

FIGURE 9. Hyperrotated Recurrent Cecocolic torsion with dyspareunia

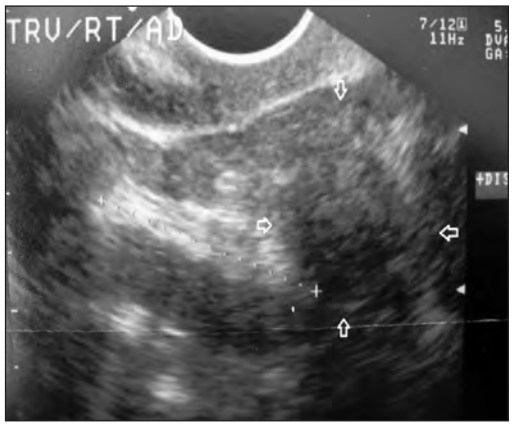

FIGURE 10. Cystic Cecocolon by ultrasound: "Phantom Tumor"

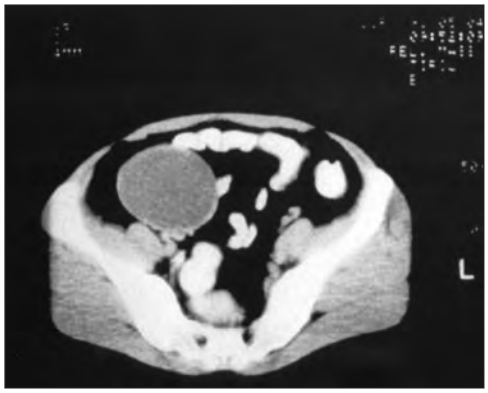

FIGURE 11. Cystic Cecocolon by CT scan: "Phantom Tumor"

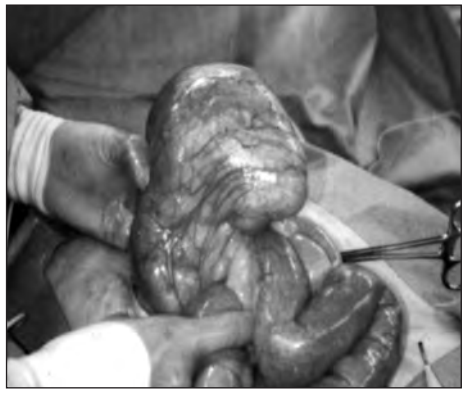

FIGURE 12. Hyperrotated Cecocolic Pseudotumor: due to subacute recurrent torsions

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entire cecocolon to the parietal peritoneal wall resulting in either a mobile cecum or a mobile cecum and ascending colon.[16] This nonfixation results in a preternatural mobility which imbues the cecocolon with the potential to torse. Jackson's membrane[13] or a Parietocolic membrane[14] may be present and act as fulcrum to the torsion. They have been mistaken for adhesions.

Intestinal torsion is the incomplete twisting of any segment of a bowel loop which may result in a partial intestinal obstruction and rarely a strangulation (Figure 6). A volvulus is a completed torsion of three hundred sixty (360) degrees and obligatorily obstructs and strangulates structurally. An algorithm[5] traces the pathogenesis of acute cecocolic torsion which may never occur because it may be preceded and interrupted by any of the other morphological variations of the intermediate forms of recurrent cecocolic torsion. These intermediate forms may be the symptomatic end-point of the per se.[5]

The normal cecum may fold upon itself but it cannot anatomically torse to form a volvulus because it is wider than long (Figure 1). Cecal volvulus, is a true misnomer because the cecum cannot anatomically twist and torse to form into volvulus (Figure 2). It has been imaginatively illustrated by some workers in the field.[1,7,10] A misleading article describe as volvulus an untorsed cecum which was strangulated by a fallopian tube.[18] Cecal bascule does not easily strangulate since the cecum has a double blood supply nor does torsion of the ascending colon because of the intact blood supply preceding and following the torsed bowel segments.

SYMPTOMS AND SIGNS

It is estimated that the majority or about seventy per cent of the patients with mobile and hyperrotated cecocolon will be asymptomatic throughout their lifetime.[3,4] Attacks of colic can start from any age into adulthood.[5,7] Acute

attacks are usually preceded by subacute episodes in seventy five percent out of one hundred cases reviewed by workers in the field.[11] When the chronic and recurrent pains becomes continuous and unrelenting, the situation has likely become an acute cecocolic torsive event.

The most common symptoms are the baffling episodic mesenteric symptom of anorexia, nausea, vague epigastric discomfort which may usher recurrent pains in the right lower abdominal quadrant. This is sometimes associated with a palpable but evanescent ballotable tumescence which has been referred to as a "phantom tumor"[19,20] (Figure 7). Recurrent bloating, constipation, and release is also complained of not unlike the symptoms of irritable bowel syndrome. Thrust dyspareunia has also been documented in either the supine or sitting attitude and is explained by the eccentric pelvic location of the cecum[5,21] (Figure 8). Currant jelly stools from admixed mucus and hemorrhagic necrotic mucosa may be noted specially among the pediatric population[5,23] (Figure 3). If plain abdominal x-rays have been performed, the radiologist may detect and should report an eccentric cecal shadow dislocated away from the right iliac fossa (Figure 9).

The most significant history is from the patient who has had many tests including a negative colonoscopy and gynecological examination. Xrays may have been taken but without any reported positive findings. This does not void the suspicion of an unreported mobile or hyperrotated cecocolon, so the films need to be reviewed to confirm the absence of an eccentrically located cecal shadow.

Many undiagnosed patients are dismissed as psychologically and emotionally impaired, hypochondriacs, depressed, and some are habituated to pain medications.[19] Some have had negative appendectomy,[22] oophorectomy, salpingectomy, hysterectomy, or multiple lower abdominal and pelvic surgeries without relief of symptoms. Many have been dismissed after a negative colonoscopy which, in this instance, is never diagnostic.[5,20]

The patient may have had a palpable cystic mass by internal examinations which may or may not be confirmed by another examiner or a sonogram disclosing the cystic mass but at another date a repeated sonogram is negative (Figure 10). Even more puzzling is when at surgery the palpated mass is not found, a veritable phantom tumor.

RADIOLOGY

Positional contrast enema is the single most accurate diagnostic examination that will disclose a hyperrotated, elongated, and mobile cecocolon. It can also exquisitely define and differentiate it from a normally rotated and mobile cecum or cecocolon.[23] Barium is preferable to water soluble contrast enema unless there is a concern about producing a colonic leak.[23]

A plain film of the abdomen may suffice to reveal the eccentrically mobile cecocolon located away from the iliac fossa. Ultra sonography can disclose the cystic configuration of a recurrent cecocolic torsion (Figure 10). The CT scan can disclose the same finding (Figure 11) and if present, will confirm the presence of a cecocolic pseudotumor[10] (Figure 12).

The radiologist should report all encounters with an eccentrically located cecum or cecocolon, even if it is seen per happenstance, because of its potential to be symptomatic. The congenitally hyperrotated cecocolon being elongated, can form into a pseudocyst[5,23] (Figure 10) or a phantom tumor[19,20] (Figure 11). A pseudotumor may develop because

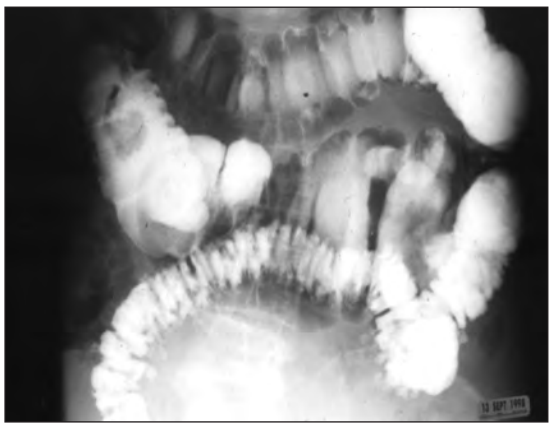

FIGURE 13. Hyperrotated Elongated Cecocolon incarcerated as left inguinal hernia

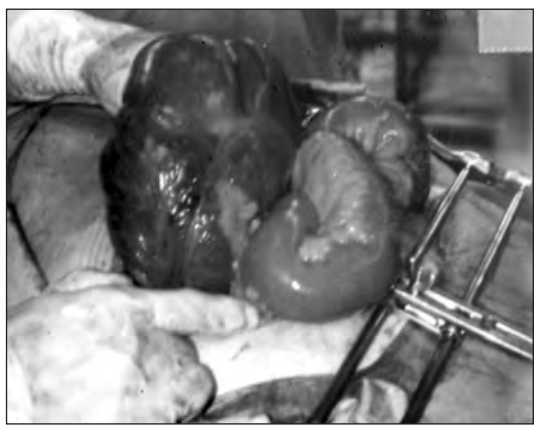

FIGURE 14. Acute Cecocolic Torsion: still viable and treated with cecopexy and cecostomy

of chronic recurrent attacks of obstruction producing in essence thickened hypertrophied walls[5,14] (Figure 12). It may incarcerate as a left inguinal hernia, (Figure 13) have the appendicitis occur in the inguinal canal,[24] locate in the true pelvis and cause dyspareunia[5,21] (Figure 8). It may be the etiology of some of the avowed irritable bowel syndrome.[25] It can be misnamed as cecal volvulus[1,10] (Figure 14). A question that begs an answer is whether the pseudocystic configuration is actually the true etiology of Ogilvie's syndrome of pseudo-obstruction[14] (Figure 6).

During an ongoing attack of torsion-obstruction, a plain film of the abdomen may disclose the gas or air-fluid filled cecocolon with a classical tapering gas shadow called the birds beak sign.[2] There may not be air-fluid filled loops of bowel proximal to the site of torsion nor collapsed loops of bowel distal to the obstruction[1,23,26] because these are basically partial obstructions (Figure 6).

The "whirl sign" by CT scan of a purported cecal volvulus is an exponential eidetic overreach with an illustration that purports to show the lines of a completed three hundred sixty degree torsion of the ileocecocolon and its mesenteric and mesocolic root. If this is true, it should be more representative in ovarian, midgut, or sigmoid volvulus to cite the few organs that may form a volvulus. It can not be pathognomonic of "cecal volvulus," by any hypothetical extension, because this entity is nonexistent. This illustration could have been confirmed and verified quite simply at operation as a three hundred (360) degree completed torsion, given that currently there is a plethora of surgical photographic availability.[1] Moreover if the "whirl lines" as seen need not be necessarily lines of volvulus but could be lines of torsion as well, which would still be germane to alert the radiologist of an ongoing pathological process.

DIAGNOSIS

The history of vague mesenteric symptoms, undiagnosed recurrent right lower abdominal quadrant pain, evanescent ballotable tumescence, thrust dyspareunia, negative colonoscopy, negative appendectomy, and negative abdominal explorations should alert the clinician to screen the patient with a the plain film of the abdomen. If it has already been obtained, it should be reviewed for an eccentric cecal shadow which is away from the right iliac fossa.

A positional contrast enema with barium or gastrografin is diagnostic. The assiduous radiologist will not need to be reminded that the active positioning by rolling the patient on the table is crucial to divulge a peripatetic cecocolon heavy with contrast medium. The relief of pain and discomfort of the patient during the procedural positioning is in itself diagnostic and therapeutic by the detorsion of a kinked bowel.

COMMENT

The congenital hyperrotated cecocolon as a clinical entity lies below the level of diagnostic awareness. It is germane to formalize the diagnosis because there are many symptomatic but undiagnosed suffering patients. The diagnostic failures are a result many causes; foremost are: meager treatises, didactic misdirection, and inaccuracy of those published. The habitual dependence and reliance on empirical inferences, and on conclusions by consensus reinforces this default. The copious photographic documentation presented should validate this diagnosis.

TREATMENT

Right hemicolectomy or segmental ileocolectomy with ileocolostomy is the most definitive surgical treatment for the symptomatic hyperrotated and elongated cecocolon. It is advisable to perform cecocolopexy of a residual mobile right colon to the parietal peritoneal wall.[5] Detorsion by insufflations or contrast enema is at best palliative and temporary.

CONCLUSION

The congenital hyperrotated and elongated cecocolon is a proved clinical entity. Because it is preternaturally mobile it has the propensity to torse, obstruct, and rarely, to strangulate. It is estimated to occur in ten to twenty per cent of the population who has cecocolic mobility. The chronic recurrent cecocolic torsion with spontaneous resolution is the most common variant. This subacute form may proceed into an acute torsion and obstruction with or without strangulation.

The symptoms are protean and should be sought in any patient with baffling mesenteric symptoms which is usually associated with right lower abdominal pains and gynecological complaints. Colonoscopy is not diagnostic.

A plain film of the abdomen may reveal the elongated cecocolon showing the cecum located away from the right iliac fossa. Definitive diagnosis is confirmed by positional contrast enema. The radiologist should be report any eccentrically located cecocolon even if seen per happenstance. An ultrasound may disclose a cystic configuration of the cecocolic torsion. The definitive treatment is a right hemicolectomy.

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