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Radiologic and Morphodynamic Profiling of the Achalasia Subtypes Described in the Chicago Classification V4.0

Fontanella G[1]*, Borrelli S[2], Brogna B[3], Parente E[4], Russo M[5], Fuggi G[6], Bianchi L[7], Arturi A[8] and Manganiello CAT[9]

[1]Department of Radiology, Ospedale Sacro Cuore di Gesù FBF, Benevento, Italy

[2]Hillman Cancer Center Villa Maria, Mirabella Eclano, Italy

[3]Department of Radiology, A.O.R.N. 'S.G.Moscati', Avellino, Italy

[4]Department of Surgery, Ospedale Sacro Cuore di Gesù FBF, Benevento, Italy

[5]Department of Surgery, Ospedale Sacro Cuore di Gesù FBF, Benevento, Italy

[6]Department of Gastroenterology, Ospedale Sacro Cuore di Gesù FBF, Benevento, Italy

[7]Department of Radiology, Ospedale Sacro Cuore di Gesù FBF, Benevento, Italy

[8]Head of Department of Surgery, Ospedale Sacro Cuore di Gesù FBF, Benevento, Italy

[9]Head of Department of Radiology, Ospedale Sacro Cuore di Gesù FBF, Benevento, Italy

***Corresponding author:**

Giovanni Fontanella,
Department of Radiology, Ospedale Sacro Cuore di Gesù FBF, Benevento, Italy

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1. Introduction

The need for a new, systematic way of approaching, interpreting and reporting barium swallow, especially in patients with achalasia, arises from the fact that we believe that in life and in medicine in particular, there should not be waste of resources, knowledge and time. Too many a time we have witnessed poorly conducted examinations with sub-par reporting. Achalasia may be a rare disease, but a radiologist should always be aware of the possibility of facing a patient with dysphagia and should be fully prepared to carefully execute and correctly interpret the images. As stated before, the reporting process begins while taking the clinical history, when usually the patient, especially when wisely guided by the physician, points out his problems very clearly. The actual reporting does actually continue while the exam is being executed, with images being interpreted real-time, in-vivo, allowing the imaging protocol to be tailored on the specific needs of the patient and, of course, of the radiologist. The FBF Scoring System [1] (FBF being the Fatebenefratelli Hospital in Benevento, Italy, where we are based and the scoring system was developed) is not only just a way of prognostically dividing patients in three subtypes, in complete agreement to clinical phenotypes cited in the manometric Chicago Classification [2]; it is rather an organic and systematic pathway that guides the radiologist, using a simple checklist, from image interpretation to structured reporting, avoiding easy mistakes and upgrading the overall quality of the service. The FBF scoring system is the first systematic radiological classification not to be based just on morphology alone, specifically varying degrees of esophageal dilatation, because it integrates dynamic findings with morphological data, making it rather improbable to miss a diagnosis. The most important aspect of the FBF scoring system, in our opinion, is that being completely in agreement to the clinical-manometric Chicago Classification, radiologists, gastroenterologists and surgeons and the rest of the Upper GI Multidisciplinary Team end up speaking the same 'language', this meaning that a radiology report will be finally something really worth reading and relying upon, a great support to the endoscopic and, especially, manometric diagnoses. At the same time, the simple FBF checklist breaks the image interpretation process down to just five parameters to assess and is the direct result of the profiling study of the disease we are presenting hereby, a preliminary study we conducted before the FBF Score one.

2. The FBF Checklist

The FBF checklist, as shown in (Table 1), is a simple list of five imaging findings, or parameters, that have to be looked for and assessed, in order to correctly diagnose and grade achalasia patients. These findings, namely bird-beak sign, esophageal dilatation, hypotonia, endoluminal stasis and spasm, are those to look upon to effectively interpret barium swallow in a patient with achalasia [3]. Each of these findings is given a specific score, when present. The different combinations of present findings, with the aid of the checklist and scoring system, the FBF Score we developed, are efficiently linked to a specific achalasia subtype.

FBF CHECKLIST
Bird-beak sign
Esophageal Dilatation
Hypotonia
Endoluminal stasis
Spasm

Table 1

3. Bird-Beak Sign

The bird-beak sign (Figure 1), sometimes even referred to as 'rat-tail sign' is maybe the best known and most easily recognisable radiographic sign of achalasia. The term bird-beak refers to the shape of the tapered, conical and smooth esophageal lumen in proximity of the gastro-oesophageal junction, that is significantly narrowed in patients with achalasia. While this sign is common to all three achalasia subtypes, it is more easily recognised in patients with classic, hypotonic subtype 1 achalasia [4]. The involved esophageal segment is generally 1 to 3 cm long, with smooth mucosal profile, with no sign of abrupt contour changes, local masses nor nodularity; some degree of pliability is retained. Differential diagnosis mainly includes intrinsic or extrinsic malignancies, but other conditions, too, such as peptic strictures, Chagas' disease, pancreatic pseudocysts, post-surgical complications (especially after fundoplicatio and vagotomy). Findings suspect for secondary achalasia are many. A tapered segment long at least 3.5 cm or more is highly suspect for regional malignancy, especially cancer of the gastric cardia and esophageal adenocarcinoma. Pseudo- or secondary achalasia are generally characterised by a longer, rigid, asymmetric and irregularly shaped tapering; sometimes, especially in the the case of malignancy of the cardia, the involved segment might include the distal esophagus even above the gastroesophageal junction. In order to correctly identify a bird-beak sign, we invite the reporting physician to check the presence of all the items in (Table 2).

BIRD-BEAK SIGN
Smooth, conical tapering
Length of EGJ involved: 1-3 cm
Relatively symmetrical
No nodularity
No signs of local masses

Table 2

4. Esophageal Dilation

In 1980, in his seminal work Gastrointestinal Radiology, Marcel Brombart described achalasia as single, progressively deteriorating condition, in which, apart from the classic bird-beak sign, the main factor to keep into consideration was lumen dilation, describing four stages of the disease: Stage I, mild achalasia, diagnosed when lumen caliber measured less than 4 cm (Figure 2); Stage II, or moderate achalasia, in which the caliber of the oesophageal lumen measured between 4 and 6 cm (Figure 3); Stage III, or severe achalasia, diagnosed when the caliber measured 6 cm or more (Figure 4). Sigmoid achalasia, in which the overtly dilated, unpropulsive lumen basically folds upon itself, is described as end-stage, or Stage IV achalasia (Figure 5). Even considering the relatively recent introduction of the Chicago Classification and all its implications in the clinical practice, these assumptions connecting lumen caliber and dilation with the severity of disease still yield some degree of truth [5], though especially when considering the evolution of the disease in singularly taken patients and, obviously, in patients with Subtype 1, hypotonic achalasia, in which the dilation is more evident than in other subtypes. It is clear, however, that even though esophageal dilation is an important parameter to assess in all patients with achalasia, because it can be present in all subtypes, albeit in varying degrees, it is just one of the many findings that characterises this disease and not the main discriminating factor between different stages. We do integrate Brombart's teachings in our morphodynamical analysis of the achalasic esophagus by keeping his dimensional staging of dilation in four degrees. Esophageal dilation is present when the caliber is at least 3 cm in diameter, considering the lower axis of the esophagus. In order to correctly identify and grade esophageal dilation, we invite the reporting physician to check all the items in (Table 3).

ESOPHAGEAL DILATION
Present when caliber > 3 cm
Grade I : caliber < 4 cm
Grade II: 4 cm < caliber < 6 cm
Grade III: caliber > 6 cm

Table 3

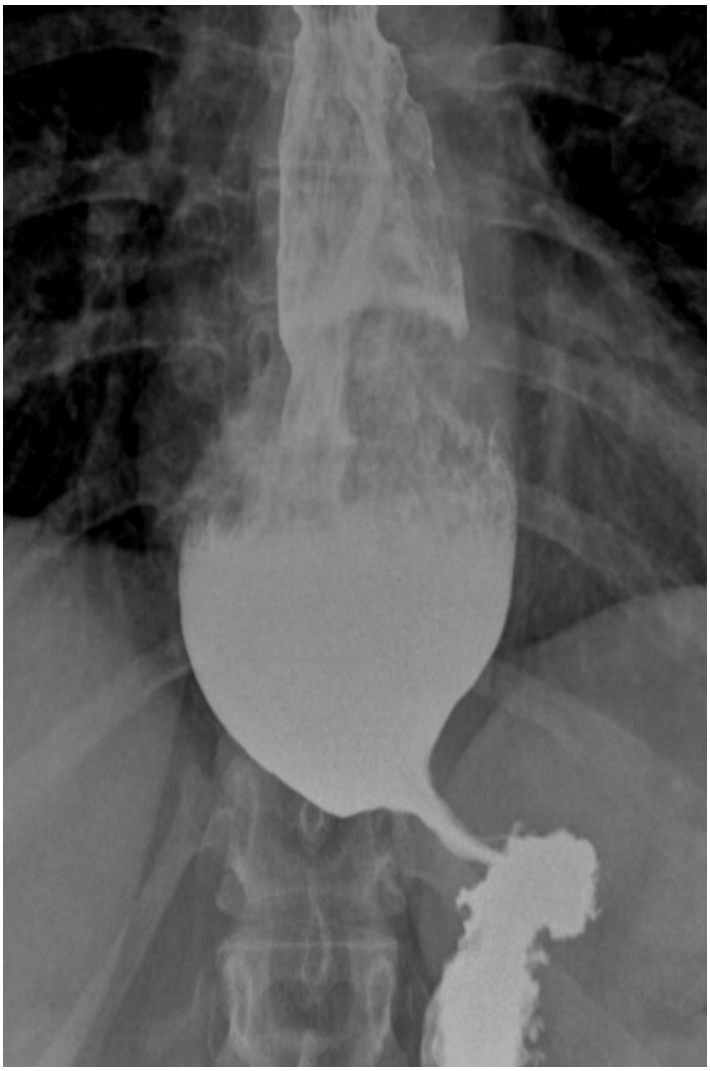

Figure 1: Bird Break Design

Figure 2: Dilation Stage I

Figure 3: Dilation Stage II

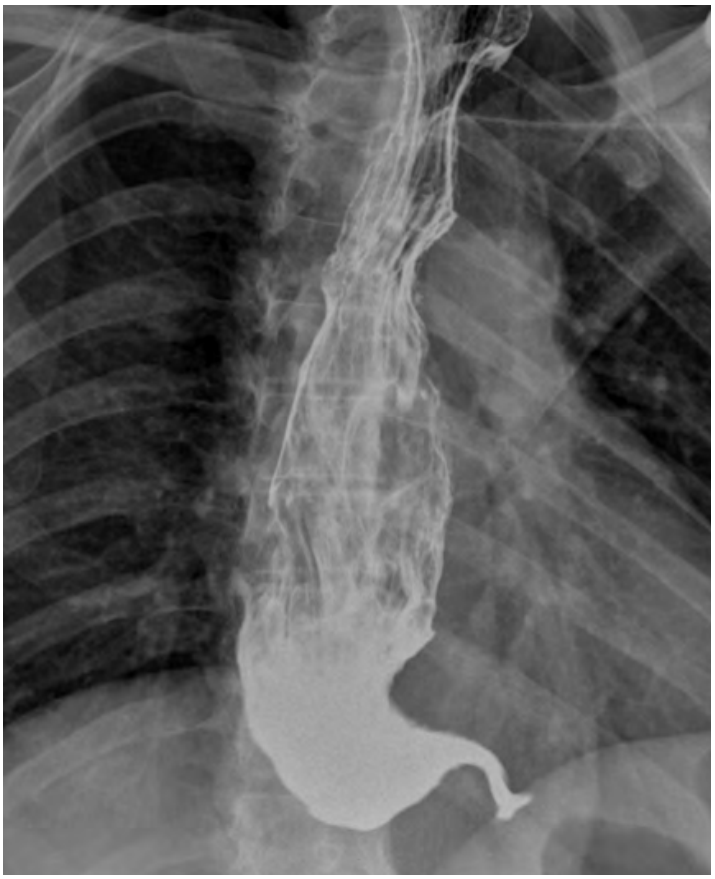

Figure 4: Dilation Stage III

Figure 5: Stage IV Sigmoid achalasia

5. Visceral Hypotonia

Hypotonia or, in some cases, atonia [6] is one of the main radiographic features of achalasia (Figure 6). To be precise, our clinically-driven approach to the morphodynamical analysis leads us to the conclusion that hypotonia, defined as absence of apparent esophageal contractions, with no episodes of panpressurization nor spasms, is actually a typical feature of Subtype 1 achalasia, often associated with esophageal dilation of varying degree. Our definition of hypotonia, deeply based on the Chicago Classification pathophysiologic assumptions, if kept in mind when reviewing dynamical barium swallow series, effectively enables the reporting physician to diagnose Subtype 1 achalasia, ruling out Subtypes 2 and 3. Rather than assessing hypotonia, what we do is actually assessing esophageal motility to recognise the exact motility pattern in the specific patient. At the same time, while the presence of spasm effectively rules out Subtypes 1 and 2, the absence of both hypotonia and spasms rules out Subtypes 1 and 3, configuring the presence of a Subtype 2, in which panpressurization is evident, with tertiary, unpropulsive waves (Figure 7); at the same time, recognising a panpressurization pattern on a barium swallow is not easy and not standardizable, that is why we developed the checklist this way and reach a Subtype 2 diagnosis with FBF Score by exclusion. This is, in our opinion, the most important pattern to be analysed, the true essence of morphodynamical imaging in achalasia; in order to effectively accomplish this assessment a thorough knowledge of pathophysiology and a basic grasp of manometry are needed. In order to correctly identify esophageal hypotonia, we invite the reporting physician to check all the items in (Table 4).

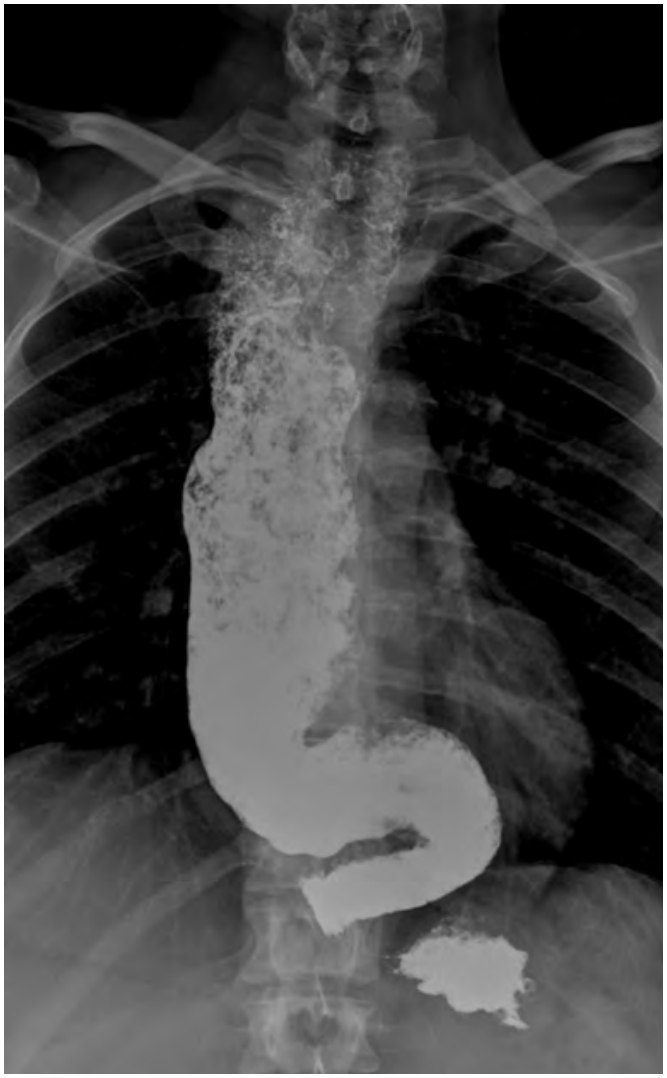

Figure 6: Hypotonia, Achalasia Subtype I

HYPOTONIA
No apparent contractions
No panpressurization
No spasms

Table 4

6. Endoluminal Stasis

Considering the intrinsic nature of achalasia, another important radiologic hallmark of all forms of the disease is endoluminal stasis. There are many ways to accomplish the assessment of barium stasis in the esophageal lumen. One way of doing this is, of course, by using a Timed Barium Esophagogram by acquiring images after the patient ingests a 200 ml bolus in 15-20 seconds, at 1, 2 and 5 minutes, subsequently tracing the upper barium level in all three images, comparing them. We tend to use this method of assessing stasis only in patients who are to undergo pneumatic dilation, or other surgical/endoscopic treatment, because it allows an effective and precise quantification of the esophageal emptying, too, enabling a comparison between pre- and post-treatment images, to check on, and quantify, eventual improvements. Now, the presence of endoluminal stasis is generally very evident, even during the execution of the exam; in order to allow correct reporting, though, efficient diagnostic criteria are needed. To assess endoluminal stasis, during the examination and after at least two boluses are ingested, we invite the patient to ingest another consistent barium bolus, a test bolus, after making sure the current barium level is under the aortic portion of the esophagus; we then proceed to acquire two short series of the same length, usually 2 or 3 seconds, 3 images per second, at 1 and 5 minutes after ingestion, considering that at 5 minutes the esophagus should be clear of endoluminal content in healthy individuals [7,8]. Upper barium levels are traced and compared (Figure 8). Barium stasis is confirmed when the heights of the barium columns are comparable (with a tolerance margin of 10 mm). Another way of comparing evaluations at 1 and 5 minutes might be, by tracing and calculating the areas occupied by barium. In order to correctly identify endoluminal stasis, we invite the reporting physician to check all the items in (Table 5).

ENDOLUMINAL STASIS
Evaluation at 1 and 5 mins after test bolus
Comparable height of barium columns (+- 10 mm)

Table 5

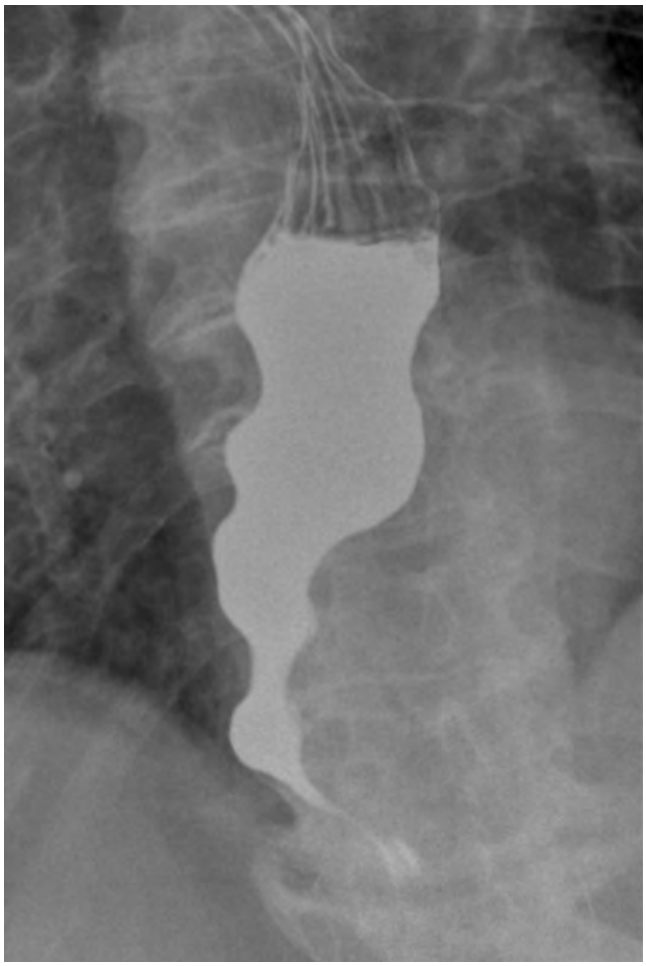

Figure 7: Panpressurization Achalasia Subtype2

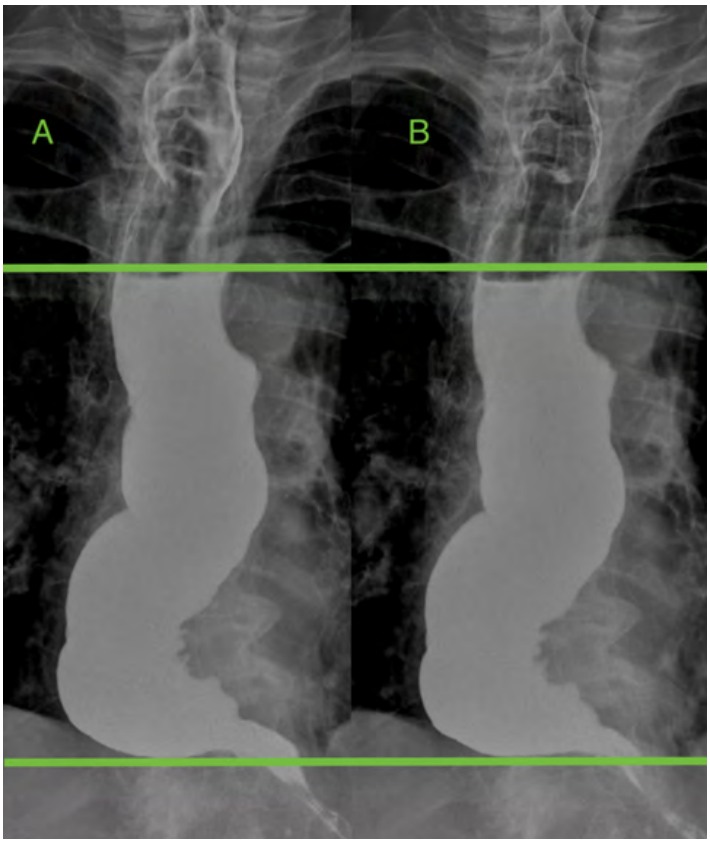

Figure 8: Luminal Stasis, Timed Barium Swallow

7. Spasm

The presence of spasm is practically pathognomonic for Subtype 3 patients, just as the presence of hypotonia/atonia is typical in Subtype 1 patients [9]. Spasm is easily detected at the morphodynamical assessment, usually in a radiographic imaging pattern already very suggestive for Subtype 3, previously known as vigorous achalasia (Figure 9). What we intend for spasm should not to be mistaken for the tertiary, uncoordinated and unpropulsive waves typical of (but not exclusive to) the panpressurizing Subtype 2 and present in the whole of the esophagus; it is rather the product of impaired relaxation of the LES plus active spastic contractions of the mid- and distal esophagus that translate into what is known as a corkscrew pattern, an irregular succession of massively narrow and dilated esophageal portions. This is what differentiates pressure disorders in Subtypes 2 and 3; they are both unpropulsive, but in Subtype 2 we witness a rather moderately dilated lumen with sporadical tertiary waves and pressurisation, not a succession of narrowed and dilated lumen, typical of Subtype 3. This characteristic, spastic pattern is what, most probably, makes Subtype 3 the least susceptible to treatment. Moreover, the presence of spasm is often associated with the presence of pseudodiverticula, especially epiphrenic, also seen in (Figure 9); in achalasia, this happens almost invariably in Subtype 3 patients. In order to correctly identify spasm, we invite the reporting physician to check all the items in (Table 6).

Figure 9: Spasm, Achalasia Subtype 3

SPASM
Mid-, lower esophagus, LES involved
Corkscrew pattern

Table 6

8. Conclusion

This profiling work was done as a preliminary study aimed at the detection of the essential radiologic findings to search for in patients with achalasia, the scoring items of the FBF score, taking into account the latest developments arising from the manometric assumptions of the Chicago Classification v4.0. The concept of achalasia being three different entities, with some elements in common, such as the omnipresent bird-beak sign, is crucial for the clinician approaching a dysphagic patient, because many studies that were once deemed negative or inconclusive, for technical or 'cultural' problems, now can be carefully interpreted and recognised as pathology [10,11,12].

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