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Ⅲ. 研究成果の刊行物・別冊

SPECIAL REPORT

GENERAL RULES FOR RECORDING ENDOSCOPIC FINDINGS OF ESOPHAGOGASTRIC VARICES (2ND EDITION)

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General rules for recording endoscopic findings of esophageal varices were initially proposed in 1980 and revised in 1991. These rules have widely been used in Japan and other countries. Recently, portal hypertensive gastropathy has been recognized as a distinct histological and functional entity. Endoscopic ultrasonography can clearly depict vascular structures around the esophageal wall in patients with portal hypertension. Owing to progress in medicine, we have updated and slightly modified the former rules. The revised rules are simpler and more straightforward than the former rules and include newly recognized findings of portal hypertensive gastropathy and a new classification for endoscopic ultrasonographic findings.

Key words: endoscopic findings, esophagogastric varices, portal hypertension.

INTRODUCTION

Bleeding from esophagogastric varices is a catastrophic complication of chronic liver disease. A precise system for the systematic evaluation and recording of esophagogastric varices is essential for the management of portal hypertension. In 1980, the Japanese Research Society for Portal Hypertension proposed a new system called 'The General Rules for Recording Endoscopic Findings on Esophageal Varices'.1 In 1991, the revised rules were proposed as 'General Rules for Recording Endoscopic Findings of Esophagogastric Varices (1991).^{2,3} These rules have gradually been accepted and are now used in many countries. Recently, portal hypertensive gastropathy (PHG) has been recognized as a distinct histological and functional entity. Endoscopic ultrasonography (EUS) can now clearly depict vascular structures around the esophageal wall in patients with portal hypertension. Owing to such progress in medicine, we slightly revised the former rules. The revised rules are simpler and more straightforward than the former rules, and include newly recognized findings of PHG and a new classification for endoscopic ultrasonographic findings.4

OUTLINES OF THE NEW RULES

The revised rules are titled 'General Rules for Recording Endoscopic Findings of Esophagogastric Varices (2nd Edition)'. These new general rules incorporate recommenda-

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tions for recording newly recognized findings of PHG and a new classification for findings on EUS. Similar to the former rules, the revised rules comprise six main categories: location (L), form (F), color (C), red color signs (RC), bleeding signs, and mucosal findings (Table 1). In principle, the endoscopic diagnosis is based on endoscopic findings assessed with the naked eye. Findings of gastric varices, which were included with esophageal varices in the former rules, are now listed separately.

Endoscopy

Esophageal varices

Location. There is no change in the rules for location (L). The longitudinal location of esophageal varices (EV) of different caliber is classified into three distinct regions: (i) locus superior (Ls) varices are located in the upper part of the esophagus; (ii) locus medialis (Lm) varices are located in the middle part of the esophagus; and (iii) locus inferior (Li) varices are located in the lower part of the esophagus.

Form. The rules for form (F) also remain the same. EV are classified into four groups according to their shape and size: (i) F_0 lesions lack a varicose appearance. This classification is useful for documenting the disappearance of EV in response to treatment, even if red veins or blue veins are present (Fig. 1a,b); (ii) F_1 lesions are straight, small-caliber varices. Small venous dilatations that disappear on insufflation of the esophagus are not included in this subgroup (Fig. 1c); (iii) F_2 lesions are moderately enlarged, beady varices (Fig. 1d); and (iv) F_3 lesions are markedly enlarged, nodular or tumor-shaped varices (Fig. 1e).

Table 1. General rules for recording endoscopic findings of esophagogastric varices (2004)

Category	Code subcategory
Location	Ls: Locus superior
(L)	Lm: Locus medialis
	Li: Locus inferior
	Lg-c: Adjacent to the cardiac orifice
	Lg-cf: Extension from the cardiac orifice to the fornix
	Lg-f: Isolated in the fornix
	Lg-b: Located in the gastric body
	Lg-a: Located in the gastric antrum
Form (F)	F ₀ : No varicose appearance
()	F ₁ : Straight, small-caliber varices
	F ₂ : Moderately enlarged, beady varices
	F ₃ : Markedly enlarged, nodular or tumor-shaped varices
Color (C)	Cw: White varices
	Cb: Blue varices
	Cw-Th: Thrombosed white varices
	Cb-Th: Thrombosed blue varices
Red color	RWM: Red wale markings
signs	CRS: Cherry red spots
(RC)	HCS: Hematocystic spots
	Esophageal varices: RC ₀ , RC ₁ , RC ₂ , RC ₃
	Gastric varices: RC ₀ , RC ₁
	Te: Telangiectasia
Bleeding	Gushing bleeding
signs	Spurting bleeding
	Oozing bleeding
	Red plug
	White plug
Mucosal	E: Erosion
findings	Ul: Ulcer
	S: Scar

Color. The color of EV was referred to as 'fundamental color' in the former rules. In the revised rules, the 'color' of EV is classified into two categories: (i) white varices (Cw) are whitish and look like large folds of the esophageal mucosa (Fig. 2a); (ii) blue varices (Cb) are bluish-white or cyanotic, and distended by blood (Fig. 2b). The esophageal mucosa overlying varices of this category appears very thin. High-risk blue varices are characterized by a fully expanded appearance with a glossy surface, similar to that of an over-inflated balloon. Blue varices that have become purple or violet because of increased variceal pressure are designated as such by adding a 'v' (violet) to their color (Cbv) in the revised rules. Thrombosed varices are indicated by adding Th (thrombosis) to their color (i.e. Cb-Th [Fig. 2c] or Cw-Th [Fig. 2d]).

Red color signs. Red color signs (RC) refer to reddish changes seen immediately beneath the submucosa. RC are known to be reliable predictors of the risk of variceal bleeding⁵ and are classified into the following three categories: (i) red wale markings (RWM) are dilated venules oriented longitudinally on the mucosal surface, somewhat like wale or whip marks (Fig. 3a); (ii) cherry-red spots (CRS) are small red spots on the mucosal surface (Fig. 3b); and (iii) hemato-

cytic spots (HCS) are large, round, crimson-red projections that look like blood blisters (Fig. 3c). RC are the most important predictor of variceal bleeding.

RC are graded as 0, 1, 2, or 3 according to their density and distribution: (i) RC₀ = absent; (ii) RC₁ = small in number and localized (Fig. 3d); (iii) RC₂ = intermediate between RC₁ and RC₃ (Fig. 3e); and (iv) RC₃ = large in number and circumferential (Fig. 3f). The RC category (RWM, CRS and/or HCS) is stated in parentheses after the RC grade. RC in patients presenting with F_0 is recorded as F_0 , RC₁₋₃. If telangiectasia (Te) is noted, its presence is recorded as Te.

Bleeding signs. Bleeding signs are divided into those found during bleeding and those found after hemostasis. In the former rules, bleeding was classified as spurting or oozing. In the revised rules, bleeding is classified as gushing (Fig. 4a), spurting (Fig. 4b), or oozing. Findings after hemostasis are classified as red plug (Fig. 4c) or white plug (Fig. 4d), similar to the former rules.

Mucosal findings. There is no change in the rules for recording mucosal findings. Mucosal findings are classified into three categories: (i) erosion (E) (Fig. 5a); (ii) ulcer (Ul) (Fig. 5b); and (iii) scar (S) (Fig. 5c). The presence of these findings is recorded as E, Ul, and S.

Evaluation of the effectiveness of treatment

'Eradication' means the disappearance of varices after treatment, including thrombosed varices (F_0 , RC₀). 'Residue' means residual varices with F or RC after treatment. 'Recurrence' means the reappearance of eradicated varices (F_0 , RC₀) with F and/or RC. 'Relapse' means the worsening of residual varices with F and/or RC.

Note. Findings of EV should be recorded in the order of the six main categories (L, F, C, RC, bleeding signs, and mucosal findings) as shown in the following examples.

- 1. EV with RWM and CRS: Ls, F_3 , Cb, RC₃ (RWM, CRS).
- 2. Spurting bleeding from EV: Lm, F_2 , Cb, RC₁ (CRS), spurting bleeding.
- 3. Thrombosed blue varices treated by endoscopic injection sclerotherapy: Lm, F_2 , Cb-Th, RC₀, Ul.
 - 4. Completely eradicated EV: F₀, RC₀.
 - 5. Recurrent EV with RWM: Li, F₁, RC₁ (RWM).

Gastric varices

Location. Gastric varices (GV) are classified into three main groups based on their relation to the cardiac orifice. GV are classified as Lg-c if they are adjacent to the cardiac orifice (Fig. 6a), Lg-cf if they extend from the cardiac orifice to the fornix (Fig. 6b), and Lg-f if they are localized to the fornix (Fig. 6c). In addition, GV located in the body of the stomach are classified as Lg-b, and those located in the antrum are classified as Lg-a. This classification is based on the relation between the location and the blood supply route of GV.⁶

Red color signs. RC are graded as 0 or 1: (i) RC_0 = absent; (ii) RC_1 = GV with RWM, CRS, and/or HCS. All other codes used to describe EV are also used for GV.

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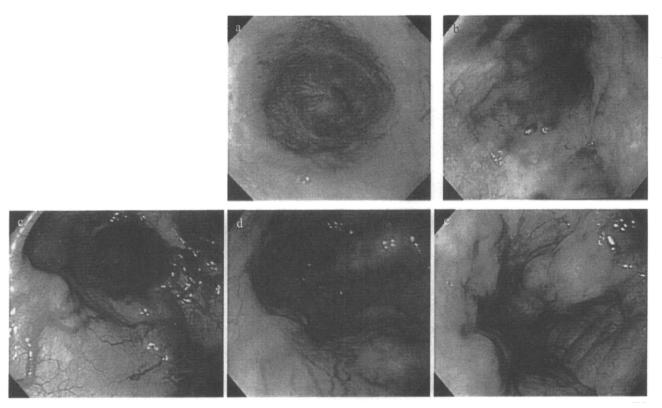


Fig. 1. F_0 lesions lack a varicose appearance. (a) F_0 , RC₀, S. (b) F_0 , RC₁. (c) F_1 lesions are straight, small-caliber varices. (d) F_2 lesions are moderately enlarged. (e) F_3 lesions are markedly enlarged, nodular or tumor-shaped varices.

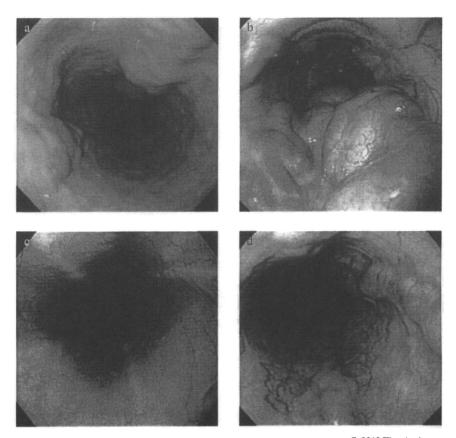


Fig. 2. (a) White varices (Cw), (b) blue varices (Cb), (c) thrombosed white varices (Cw-Th), (d) thrombosed blue varices (Cb-Th).

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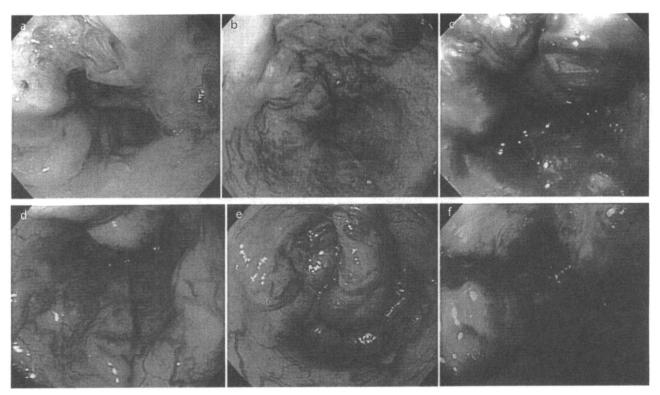


Fig. 3. Red color signs. (a) Red wale markings (RWM), (b) cherry-red spots (CRS), (c) hematocytic spots (HCS), (d) RC₁, (e) RC₂, (f) RC₃.

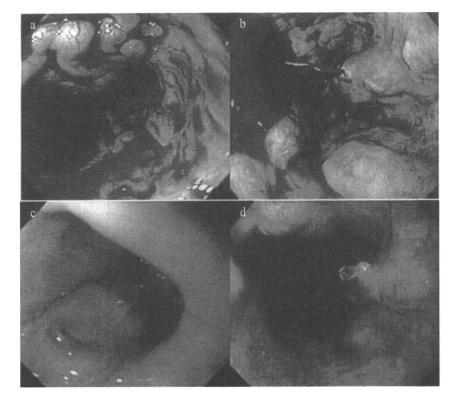


Fig. 4. Bleeding is classified as (a) gushing, (b) spurting, or oozing. Signs appearing after hemostasis are classified as (c) red plug or (d) white plug.

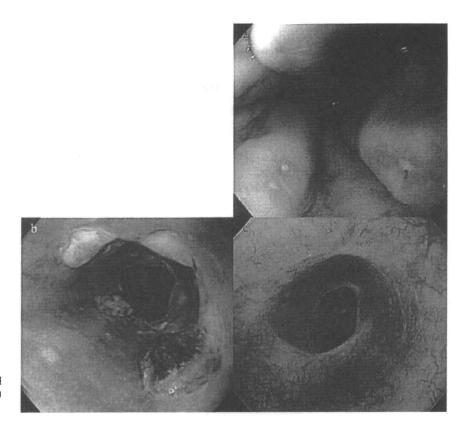


Fig. 5. Mucosal findings are classified as (a) erosion (E), (b) ulcer (UI), or (c) scar (S).

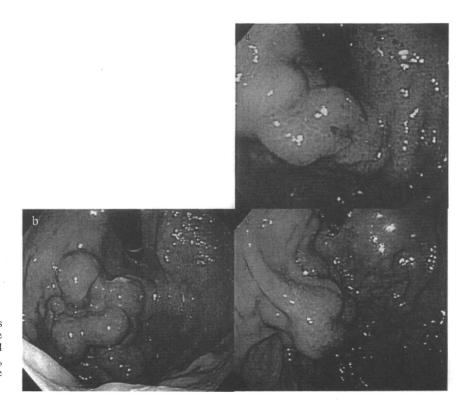


Fig. 6. Gastric varices are classified as (a) Lg-c if they are adjacent to the cardiac orifice, (b) Lg-cf if they extend from the cardiac orifice to the fornix, and (c) Lg-f if they are distant from the cardiac orifice.

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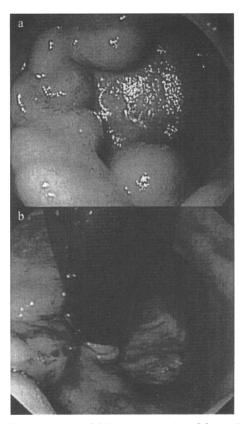


Fig. 7. Ectopic varices. (a) Duodenal varices, (b) rectal varices.

Note. Findings of GV should be recorded in the order of the six main categories (L, F, C, RC, bleeding signs, and mucosal findings) as shown in the following examples.

- 1. EV and fundic varices: Ls, F_3 , Cb, RC₃ (RWM, CRS), Te, Lg-f, F_2 , RC₀.
- 2. Spurting bleeding from GV extending from the cardiac orifice to the fornix: Lg-cf, F_3 , spurting bleeding.
 - 3. Thrombosed blue fundic varices: Lg-f, F2, Cb-Th, RC0.
 - 4. Recurrent fundic varices: Lg-f, F1, RC1.

Ectopic varices

Ectopic varices are defined as gastrointestinal varices other than esophagogastric varices. All codes for EV are used to describe ectopic varices, such as duodenal varices (Fig. 7a), jejunoileal varices, colonic varices, and rectal varices (Fig. 7b).

Portal hypertensive gastropathy

In patients with portal hypertension, bleeding can be associated with gastric mucosal lesions such as hemorrhagic gastritis, acute gastric erosions, bleeding gastritis, or acute erosive gastritis. Recently, mucosal lesions associated with portal hypertension have been referred to as PHG.⁷ Findings associated with PHG are classified into three categories: (i) grade 1, erythematous flecks or maculae; (ii) grade 2, red spots and/or diffuse redness; and (iii) grade 3,

intramucosal or luminal hemorrhage. A snakeskin (mosaic) appearance can be associated with all three grades of PHG (Fig. 8a–f).

Endoscopic ultrasonography

Esophageal varices

On EUS images, EV appear as an echo-free or hypoechoic lumen in the esophageal submucosa (Figs 9, 10a,b).

Diameter. The maximum minor-axis diameter (mm) of EV should be recorded. The absence of an echo-free lumen and hypoechoic lumen after treatment is recorded as D(0).

Perforating veins. The presence or absence of perforating veins (Pv) should be recorded as Pv(+) and Pv(-), respectively. If Pv is present, its maximum diameter (mm) should be recorded.

Peri-esophageal veins. Peri-esophageal veins (Peri-v) refer to a group of small vessels adjacent to the muscularis externa of the esophagus or partly invading the muscular wall of the esophagus. The presence or absence of Peri-v should be recorded as Peri-v(+) and Peri-v(-), respectively.

Para-esophageal veins. Para-esophageal veins (Para-v) refer to a group of rather large vessels distal to the muscularis externa of the esophagus. The presence or absence should be recorded as Para-v(+) and Para-v(-), respectively.

Note. EUS findings on EV should be recorded in the order of the four main categories (D, Pv, Peri-v, Para-v) as shown in the following examples.

- 1. EV (EUS): D (3), Pv(-), Peri-v(-), Para-v(-).
- 2. EV (EUS): D (4), Pv(+) (3), Peri-v(+), Para-v(-).

Post-treatment EUS findings. Esophageal wall thickness (mm) should be recorded as an index of hypertrophy (Hy).

Gastric varices

On EUS images, GV appear as an echo-free or hypoechoic lumen in the gastric submucosa (Fig. 10c,d).

Diameter. The maximum minor-axis diameter of GV (mm) should be recorded. The absence of an echo-free lumen and hypoechoic lumen after treatment is recorded as D(0).

Perforating veins. The presence or absence of Pv should be recorded as Pv(+) and Pv (-), respectively. If Pv is present, its maximum diameter (mm) should be recorded.

Peri-gastric veins. Peri-gastric veins (Peri-v) refer to a group of small vessels adjacent to the gastric wall or partly invading the muscular wall of the stomach. The presence or absence of Peri-v should be recorded as Peri-v(+) and Peri-v(-), respectively.

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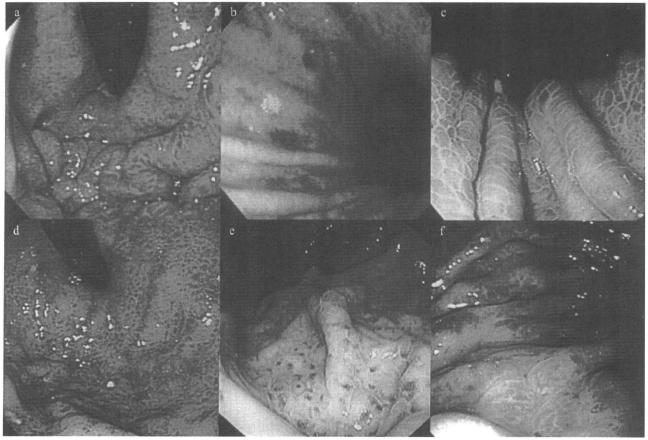


Fig. 8. (a) Fine pink speckling, (b) superficial reddening, (c) snakeskin (mosaic) appearance, (d) diffuse redness, (e) cherry red spots, (f) intramucosal hemorrhage.

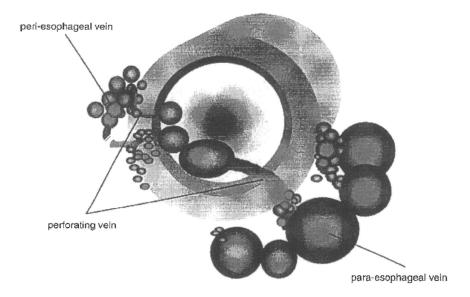


Fig. 9. Schematic drawing of the vascular structure inside and outside the esophageal wall.

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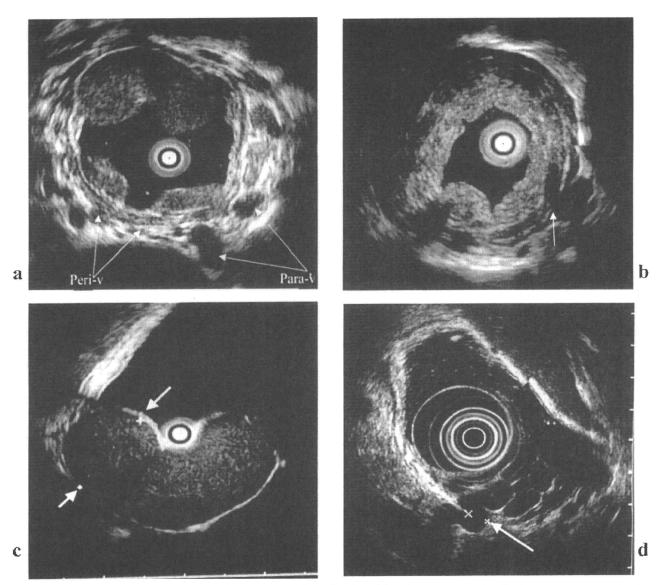


Fig. 10. Endoscopic ultrasonography (EUS) images of esophageal varices: (a) peri-esophageal veins (Peri-v), para-esophageal veins (Para-v); (b) perforating vein (Pv) (arrow). EUS images of gastric varices: (c) using a 20-MHz probe (arrow: diameter of gastric varices), (d) using a 7.5-MHz probe (arrow: perforating vein).

Para-gastric veins. Para-gastric veins (Para-v) refer to a group of rather large vessels distal to the gastric wall. The presence or absence of Para-v should be recorded as Para-v(+) and Para-v(-), respectively.

Note. EUS findings on GV should be recorded in the order of the four main categories (D, Pv, Peri-v, Para-v) as shown in the following examples.

- GV (EUS): D (8), Pv(+) (7), Peri-v(+), Para-v(+).
 GV (EUS): D (0), Pv(-), Peri-v(-), Para-v(+).

Post-treatment EUS findings. Gastric wall thickness (mm) should be recorded as an index of hypertrophy (Hy).

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