

表 2 Marfan 症候群 (MFS) 一新基準一

家族歴がない場合：	
(1) Ao ($Z \geq 2$) + EL = MFS*	
(2) Ao ($Z \geq 2$) + FBN1 = MFS	
(3) Ao ($Z \geq 2$) + Syst (≥ 7 pts) = MFS*	
(4) EL + FBN1 (大動脈拡張を伴うことが報告されている異常) = MFS	
家族歴がある場合 (すなわち上記を満たす同胞を有する)：	
(5) EL + FH = MFS	
(6) Syst (≥ 7 pts) + FH = MFS*	
(7) Ao (20 歳以上の場合 $Z \geq 2$, 20 歳未満の場合 $Z \geq 3$) + FH = MFS*	
EL + FBN1 (ただし大動脈拡張との関連が過去に報告されていない) = ELS	
Ao ($Z < 2$) + Syst (≥ 5 , 1 つ以上の骨格系異常を含む) + EL がない = MASS	
MVP + Ao ($Z < 2$) + Syst (< 5) + EL がない = MVPS	
* Sphrintzen-Goldberg 症候群, Loey-Dietz 症候群, 血管型 Ehlers-Danlos 症候群などの類縁疾患を示唆する身体所見を有していないことを確認する必要があり, 必要時には TGFBR1/2 や COL3A1 の検索を行う。	
Ao: バルサルバ洞の拡張, EL: 水晶体脱臼, FBN1: FBN1 の遺伝子異常, Syst: 全身評価項目, FH: MFS の家族歴, ELS: 水晶体脱臼症候群, MASS: MASS 表現型, MVPS: 僧帽弁逸脱症候群	
<全身評価項目> 20 点満点で, 7 点以上を障害ありとする	
・ 手指徴候陽性かつ母指徴候陽性	3 点 (一方のみの場合には 1 点)
・ 鳩胸	2 点 (漏斗胸あるいは胸郭非対称性の場合には 1 点)
・ 後足部変形	2 点 (扁平足のみの場合 1 点)
・ 気胸	2 点
・ 硬膜拡張	2 点
・ 寛骨臼突出	2 点
・ 重度の側弯がない場合に上腕長/前腕長比の低下, また両手を広げた長さとし長の比が 1.05 以上	1 点
・ 側弯ないしは後弯	1 点
・ 肘関節の伸展制限	1 点
・ 顔貌所見: 3 つ以上を満たす (長頭, 眼球陥凹, 眼瞼裂外下方傾斜, 頬骨低形成, 下顎後退症)	1 点
・ 皮膚線条	1 点
・ 近視 ($> 3D$, D: 近視度数)	1 点
・ 僧帽弁逸脱症 (種類は問わない)	1 点

れる。拡大してきた場合には経過観察期間を 3～6 ヶ月程度に短縮する。大動脈瘤の破裂や Stanford A 型大動脈解離は、救命のため緊急手術の適応である。大動脈瘤に対する予防的手術の適応大動脈径に関しては、血管壁の脆弱性が背景にあるため、通常の動脈硬化性大動脈瘤に比べてより小さい動脈径におい

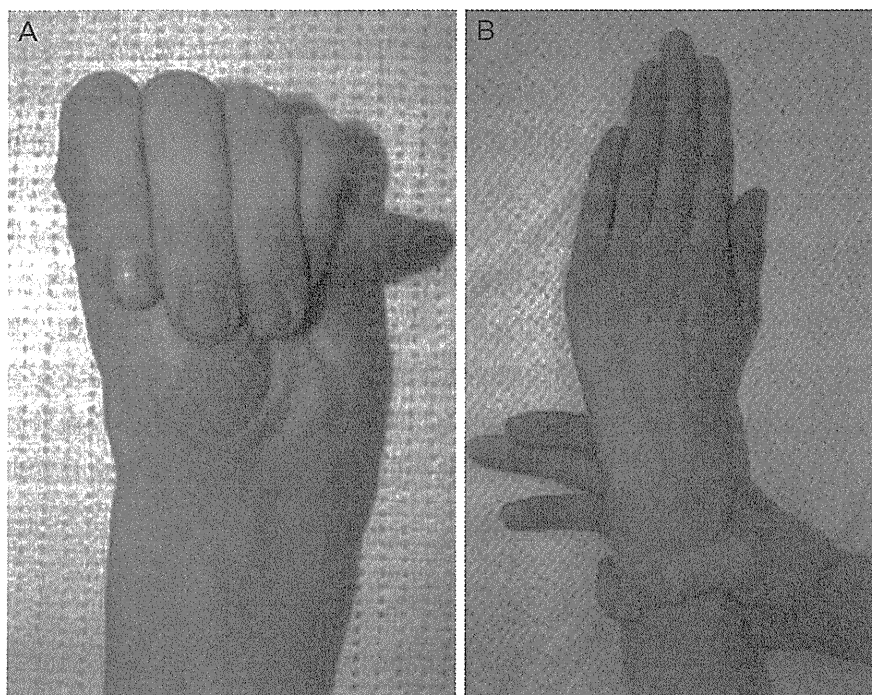
て手術が検討される。日本循環器学会の大動脈瘤・大動脈解離診療ガイドラインでは、家族歴があり上行大動脈根部が 5.0cm を超える場合 (Class IIa, Level C), 上行大動脈径が 5.5cm 以上 (Class IIa, Level C) の場合に予防的手術が推奨されている。また、上行大動脈拡張に伴い大動脈弁閉鎖不全症が進行

表3 Marfan 症候群と鑑別を要する疾患一覧

鑑別診断	責任遺伝子	特徴
心血管系異常を伴うもの： MASS 表現型	FBN1 (常染色体優性)	大動脈径拡大が軽度であり、全身所見が5点以上(少なくとも1つは骨格に伴うもの)の場合。近視 myopia, 僧帽弁逸脱症 Mitral valve prolapse, 境界域大動脈拡張 borderline Aortic root enlargement, 皮膚線条 Skin, 骨格異常 Skeletal features を主徴とする。大動脈拡張および眼症状を伴わない Marfan 症候群あるいは境界域に該当するものと思われる。経過中に大動脈拡張を生じた場合には Marfan 症候群の診断となり、定期的な心エコー検査が必要。僧帽弁逸脱症に対して、内科的治療(β遮断薬, Ca拮抗薬など)および手術療法。
僧帽弁逸脱症候群 Mitral Valve Prolapse Syndrome (MVPS)	常染色体優性, X染色体遺伝 特定の遺伝子異常は不明	僧帽弁逸脱症を有し、全身所見が5点未満の場合。僧帽弁逸脱のほか、漏斗胸, 側弯, 長く細い指などを合併。大動脈拡張および水晶体脱臼はない。僧帽弁逸脱症に対する一般的治療(β遮断薬, Ca拮抗薬などの内服および手術)。
Loeys-Dietz 症候群 (LDS) 大動脈二尖弁	TGFBR1, TGFBR2 NOTCH1, KCNJ2 など	本文参照 一部は上行大動脈瘤を合併。大動脈の拡張はバルサルバ洞と上行大動脈の接合部 (sino-tubular junction) に生じ、水晶体脱臼や骨格系異常は認めない。
Bicuspid Aortic Valve (BAV) 家族性胸部大動脈瘤症候群 (FTAA) 動脈管開存を伴う FTAA	TGFBR1/2, ACTA2 MYH11	本文参照
Ehlers-Danlos 症候群 動脈ねじれ症候群 Arterial Tortuosity Syndrome (ATS)	COL3A1, COL1A2, PLOD1 SLC2A10 (常染色体劣性)	本文参照 大動脈あるいは中動脈のねじれ, 狭窄, 瘤化, 皮膚骨格系の異常の頻度も高い。
眼異常を伴うもの： 家族性水晶体脱臼 Familial Ectopia Lentis	FBN1 (常染色体優性)	水晶体脱臼に Marfan 症候群様の骨格系異常を合併。FBN1 の遺伝子異常の場合、経時的に大動脈拡張が進行し Marfan 症候群の診断に至ることもある。
水晶体脱臼症候群 Ectopia Lentis Syndrome (ELS)	LTBP2, ADAMTSL4 (常染色体劣性)	
Weill-Marchesani 症候群 (WMS)	FBN1 (常染色体優性), ADAMTSL10 (常染色体劣性)	小球状水晶体, 短指症, 関節硬直
ホモシスチン尿症	CBS (常染色体劣性)	精神発達遅滞, 水晶体脱臼および高度近視, 骨格系異常, 血栓塞栓症など。尿中アミノ酸検査により診断が可能。
Stickler 症候群	COL2A1, COL1A1, COL11A2 (常染色体優性)	硝子体変性, 網膜剥離, 近視, 開放性隅角緑内障, 白内障など。水晶体亜脱臼はまれ。口蓋裂や難聴, 骨端異形成などを合併。Marfan 症候群様顔貌。
骨格異常を伴うもの： Sphrintzen-Goldberg 症候群 (SGS)	FBN1 異常の報告例はあるが, はっきりとは解明されていない	Marfan 症候群様体格(鳩胸あるいは漏斗胸, 側弯, クモ指症), 頭蓋顔面異常(眼球突出, 両眼解離症, 眼裂下垂, 上顎下顎低形成, 高口蓋), 発達遅滞を伴う疾患。大動脈の異常は認めない。
先天性拘縮性クモ指症 (CCA)	FBN2 (常染色体優性)	Marfan 症候群様の顔貌と長い手足が特徴。軽度の大動脈根部の拡張が認められるが進行はせず, 大動脈解離や破裂はまれ。眼症状もない。出生時に耳に皺がより上耳輪が折れ被さっていたり, 膝足関節の拘縮を認める。後側弯や関節の過伸展が認められる。

図4 Marfan 症候群における母指徴候 (A), 手指徴候 (B)

(Dean JCS: Marfan syndrome: clinical diagnosis and management.
Eur J Hum Genet 15: 724-733, 2007 より引用)



した場合にも、早期の手術を検討する。当院ではバルサルバ洞径 4.5cm 程度から手術の検討を開始している。上行大動脈を人工血管に置換する術式としては、大動脈弁人工弁置換も行う Bentall 術、自己弁を用いる David 術、Yacoub 術などがある。手術を施行しても、その後の経過で動脈拡張が進行し再手術を要する場合もある。そのため、術後の継続的な経過観察を必要とする。

Marfan 症候群の大動脈瘤に対しては、ステントグラフトは現在のところ推奨されない。

大動脈拡張の進行予防として、降圧薬による血圧コントロールが非常に重要である。従来、動脈硬化性の大動脈疾患に準じて β 遮断薬 (メトプロロール, ビソプロロール) が用いられていた。近年、アンジオテンシン II 受容体拮抗薬 (ARB) であるロサルタンが、血中 TGF β を抑制することにより Marfan 症候群において大動脈拡張の進行を遅らせると報告されており、現在臨床試験が進行中であ

る。今後、Marfan 症候群における fibrillin や TGF β の役割が明らかになるにつれ、より病態生理に直結した治療戦略が期待される。

Marfan 症候群における運動制限に関しては、患者の状態に応じて個別に判断される。高身長であるがゆえ、バスケットボールやバレーボールなどの競技で活躍していることも少なくない。一般論として、コンタクトスポーツや息こらえが必要となる等尺性の運動は制限が必要である。患者の運動耐用能によるものの、過剰負荷を強いるような激しい運動も好ましくない。軽度から中等度の有酸素運動は特に問題ない。

Marfan 症候群の女性においては、周産期 (特に妊娠後期から産褥期にかけて) に急激に大動脈径が拡大したり、大動脈解離を生じたりする可能性があるため、厳密な管理を要する。妊娠前の大動脈根部の血管径が 4.5cm 以上の場合には、周産期の大動脈拡張進行・解離発生のリスクが高まると言われ、妊娠・

出産は禁忌とされており、4.0cm 未満であれば妊娠・出産を容認するとガイドラインには記載されている。しかしながら 4.0cm 未満であっても解離、大動脈拡大を来すこともあるため、周産期には β 遮断薬で十分な降圧を図り、もし大動脈径の急激な拡大を認める場合には人工血管置換術が必要となる。

Marfan 症候群類縁疾患

先述のごとく、Marfan 症候群に関連する疾患についても病態生理や分子生物学的機序の解明が進んできた。特に鑑別診断の点で重要なものを以下に列記する。

1. Loeys-Dietz 症候群

両眼解離症、口蓋裂/口蓋垂裂、上行大動脈拡張/解離を 3 徴とする症候群であり、中にはгент基準を満たし Marfan 症候群の診断に含まれる例も認められる。常染色体優性遺伝であり、TGFBR1, TGFBR2 の遺伝子異常が同定されている。大動脈拡張/解離、僧帽弁逸脱症、側弯症、硬膜拡張など Marfan 症候群と共通の所見がある一方、Marfan 症候群では認められない頭蓋骨癒合症、Chiari 奇形、内反足変形、先天性心疾患、頸椎の不安定性、皮膚透過性亢進などが認められることで区別される。あざができやすく、皮膚は広範に癍痕化しやすい。本症候群では Marfan 症候群に比較して血管蛇行が目立ち、大動脈破裂/解離のリスクが高く、より若年かつ小さな動脈径で生じると言われており、慎重な経過観察および早期の手術介入が必要である。

2. 家族性胸部大動脈瘤 / 解離症候群

(familial thoracic aortic aneurysm and dissection syndrome)

胸部大動脈の異常を主体とした疾患群の総称であり、臨床的・遺伝的にも種々の状態が含まれている。Marfan 症候群のような

眼症状や骨格系異常は、認めないか、認めてもわずかである。原因遺伝子として FBN1, TGFBR1/2, MYH11, ACTA2 が知られている。特に後二者は、ちょうど心筋症において心筋のアクチン・ミオシン遺伝子異常が認められるのと同様、血管疾患においても血管アクチン・ミオシン異常によって血管表現型を来す点で興味深い。MYH11 の異常では動脈管開存を合併する家系が報告されており、ACTA2 の異常では虹彩異常、網状皮斑、脳動脈瘤、大動脈二尖弁、動脈管開存などを合併する。ACTA2 異常では若年性の脳血管障害や冠動脈疾患を合併することも知られている。

3. Ehlers-Danlos 症候群

複数の型が知られているが、心血管系異常を伴い、Marfan 症候群との鑑別を要するものとしては、血管型 (旧 IV 型)、後側弯型 (旧 VI 型)、古典型 (旧 I 型, II 型) の一部などが挙げられる。

血管型 Ehlers-Danlos 症候群は常染色体優性遺伝であり、III 型コラーゲン (COL3A1) の遺伝子異常が原因である。血管および組織は脆弱であり、皮膚の透過性亢進、あざができやすい、広範な皮膚癍痕形成のほか、脾・腸管・子宮破裂などの重大な合併症を生じる。中動脈レベルでの解離や破裂が典型的であるが、大動脈にも生じることがある。動脈は非常にもろく、手術に難渋することがあり、予後不良である。

後側弯型 Ehlers-Danlos 症候群は、常染色体劣性の遺伝形式をとり、後側弯、関節過伸展、筋緊張低下などの症状を呈する。大動脈の拡張および解離、中動脈の破裂を合併する。コラーゲン修飾酵素である procollagen-lysine, 1, 2-oxoglutarate 5-dioxygenase 1 (PLOD1) の酵素活性欠損が原因である。

古典型 Ehlers-Danlos 症候群は、常染色

体優性遺伝であり，皮膚や関節の過伸展および創傷治癒異常を認める．V型コラーゲンのCOL5A1, COL5A2 の遺伝子異常が原因である．古典的 Ehlers–Danlos 症候群の症例の一部において，成人後に重度の心臓弁膜症の合併が認められることがあり，心弁膜症型 Ehlers–Danlos 症候群とも呼ばれている．常染色体劣性遺伝であり，I型コラーゲンの $\text{pro}\alpha 2$ 鎖 (COL1A2) の完全欠損が報告されている．また，I型コラーゲンの $\text{pro}\alpha 1$ 鎖 (COL1A1) の遺伝子異常が認められた古典型 Ehlers–Danlos 症候群の症例において，経時的に血管型に似た表現型を認めた報告もある．

おわりに

非動脈硬化性遺伝性大動脈疾患は，その疾患分類によって表現型，予後，治療介入の手段・時期が異なるため，現在利用可能な診断

モダリティを駆使して判断することが肝要である．遺伝子診断技術は，次世代シーケンサーの開発・普及により，さらに臨床現場においてもその成果を活用する時代に入ると期待される．

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Marfan Syndrome and Related Disorders

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Is a Right Pedicle Screw Always Away From the Aorta in Scoliosis?

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Study Design. Retrospective analysis.

Objective. We evaluated the aorta safety in placement of a right pedicle screw in scoliotic patients.

Summary of Background Data. Past reports emphasized the aorta risk in placing pedicle screws on the concave left side in right thoracic scoliosis. However, risk on the right side has drawn limited interest.

Methods. Thirty-four scoliotic patients with an average age of 18.0 years were evaluated. The Cobb angle averaged $59.0^\circ \pm 14.0^\circ$. From computed tomographic data, we evaluated the aorta location relative to the spine at each level from T4 to L4 and simulated placement of a right pedicle screw with a direction different from the ideal trajectory. Sensitivity analysis was performed independently by variable direction errors and screw length: the maximum error of trajectory was set to 5° in the medial direction and to 5° , 10° , or 20° in the lateral direction, and a screw length was set at 40, 45 or 50 mm. We defined "aorta-at-risk" when a patient has some level where a simulated pedicle screw involves the aorta, and compared the curve characteristics (the apical vertebral translation, the Cobb angle and the Nash-Moe grade) between the aorta-at-risk cases and the aorta-no-risk cases.

Results. In left thoracic or lumbar curves, the aorta often resided in front of right pedicles at the periapical level. In a scenario of a simulated pedicle screw with a maximum error of 20° in the lateral direction and a screw length of 50 mm, the aorta was at risk in 7 (33%) of 21 left lumbar curves. Curve characteristics of the aorta-at-risk cases at L1 were a larger apical vertebral translation ($P = 0.003$), a larger Cobb angle ($P = 0.006$), and a larger Nash-Moe grade ($P = 0.017$) compared with those of the aorta-no-risk cases.

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Spine

Conclusion. Surgeons need to pay attention to the position of the aorta in placing a pedicle screw on the right at the periapical level of a left curve either in thoracic or lumbar spine.

Key words: scoliosis, aorta, pedicle, screw. **Spine 2011;36:E1519–E1524**

One of the disadvantages of a pedicle screw is the possibility of aorta involvement. The aorta is located on the left side of the thoracic spine and stays in front of the lumbar spine in normal subjects,¹ and past reports emphasized the aorta risk in placing a screw on the concave side, which is usually left in right thoracic scoliosis.^{2,3} On the contrary, risk on the right side in scoliosis has drawn limited interest. The purpose of this study was to evaluate the safety of the aorta in placement of a right pedicle screw in scoliosis surgery.

MATERIALS AND METHODS

A total of 34 patients with scoliosis were evaluated after excluding congenital scoliosis and soft-tissue related disease such as Marfan syndrome. The average age at a computed tomographic examination was 18.0 years (range = 10–30), and there were 4 males and 30 females. Scoliosis was idiopathic in 30 patients, Chiari-syrinx in 2, multiple epiphyseal dysplasia in 1, and Noonan syndrome in 1. There were 29 thoracic curves and 24 lumbar curves, and the Cobb angle averaged $59.0^\circ \pm 14.0^\circ$ (28° to 100° ; Table 1).

Computed tomography (CT) was taken for surgical planning with a thickness of 1.25 mm, and data were transferred to a personal computer for analysis (ExaView LITE; Ziosoft, Tokyo, Japan). In the thoracic spine, we selected the middle of the base of the right superior facet where a pedicle screw is placed as the point of origin of the coordinate system (Figure 1A). A line connecting both of the middle points of bases of the superior facets was defined as the X-axis. In the lumbar spine, we drew a line between both the medial edges of superior facets as the X-axis (Figure 1B). The Y-axis was drawn perpendicular to the X-axis starting from the dorsal edge of the right superior facet. In thoracic and lumbar spine, the angle formed by the Y-axis and a line connecting the origin and the center of the aorta was defined as the right pedicle-aorta angle, and length of a line connecting the origin and the edge of the aorta as the right pedicle-aorta distance. From the repeatability test in our

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TABLE 1. Properties of Scoliosis

ID	Diagnosis	Age (yr)	Sex	Th Side	Th Cobb (degrees)	Th Apx	L Side	L Cobb (degrees)	L Apx
8	idio	17	F	R	53	T9	n.d.	n.d.	n.d.
11	idio	19	F	R	63	T10	n.d.	n.d.	n.d.
12	idio	19	F	R	83	T9	n.d.	n.d.	n.d.
13	idio	20	F	R	51	T7	L	28	L1/2
14	idio	16	F	n.d.	n.d.	n.d.	R	50	L1
15	idio	19	F	R	57	T8	L	49	L2
19	idio	29	F	n.d.	n.d.	n.d.	L	71	L1
21	idio	12	F	R	87	T9/T10	n.d.	n.d.	n.d.
23	idio	17	F	R	57	T8	L	62	L2
24	Chiari	15	F	R	62	T4/T5	L	88	L1/2
26	idio	16	F	R	54	T7/T8	L	54	L2
27	idio	16	F	R	52	T8/T9	L	42	L2
28	idio	20	F	R	54	T7	L	37	L2
29	idio	18	M	R	50	T8/T9	L	31	L2
30	idio	18	F	R	58	T9	n.d.	n.d.	n.d.
32	idio	13	F	R	58	T8/T9	L	40	L2
33	idio	11	F	L	79	T5	R	100	L1
34	Chiari	12	M	R	52	T8/T9	L	65	L4
35	idio	29	F	R	54	T8	L	67	L2
37	idio	15	F	R	68	T9	n.d.	n.d.	n.d.
39	idio	19	F	R	57	T10	n.d.	n.d.	n.d.
40	idio	23	F	R	63	T9/T10	n.d.	n.d.	n.d.
41	idio	10	F	R	61	T10	L	35	L3
44	Noonan	23	F	R	78	T9	n.d.	n.d.	n.d.
46	idio	13	M	R	83	T9/T10	L	63	L2/3
48	MED	16	M	R	62	T9	L	42	L2/3
49	idio	21	F	n.d.	n.d.	n.d.	L	56	L1
50	idio	19	F	L	63	T5	R	64	T12
51	idio	17	F	R	66	T7	L	67	L1
53	idio	30	F	n.d.	n.d.	n.d.	L	55	L2
54	idio	13	F	R	57	T8	L	55	L2
56	idio	29	F	R	50	T7	L	55	L3
57	idio	16	F	R	58	T9	n.d.	n.d.	n.d.
58	idio	13	F	n.d.	n.d.	n.d.	L	60	L3

Dx indicates diagnosis of scoliosis; idio, idiopathic; L side, convex side of a lumbar curve; L Cobb, Cobb angle of a lumbar curve; L Apx, apex level of a lumbar curve; MED, multiple epiphyseal dysplasia; n.d., not deformed; T side, convex side of a thoracic curve; T Cobb, Cobb angle of a thoracic curve; T Apx, apex level of a thoracic curve.

previous study,⁴ interclass correlation coefficients were 0.922 to 0.957 in the intraobserver measurement and 0.896 to 0.929 (0.864–0.961) in the interobserver measurement.

We evaluated the location of the aorta relative to the spine at each level from T4 to L4 and simulated placement of a right pedicle screw with a direction different from the ideal

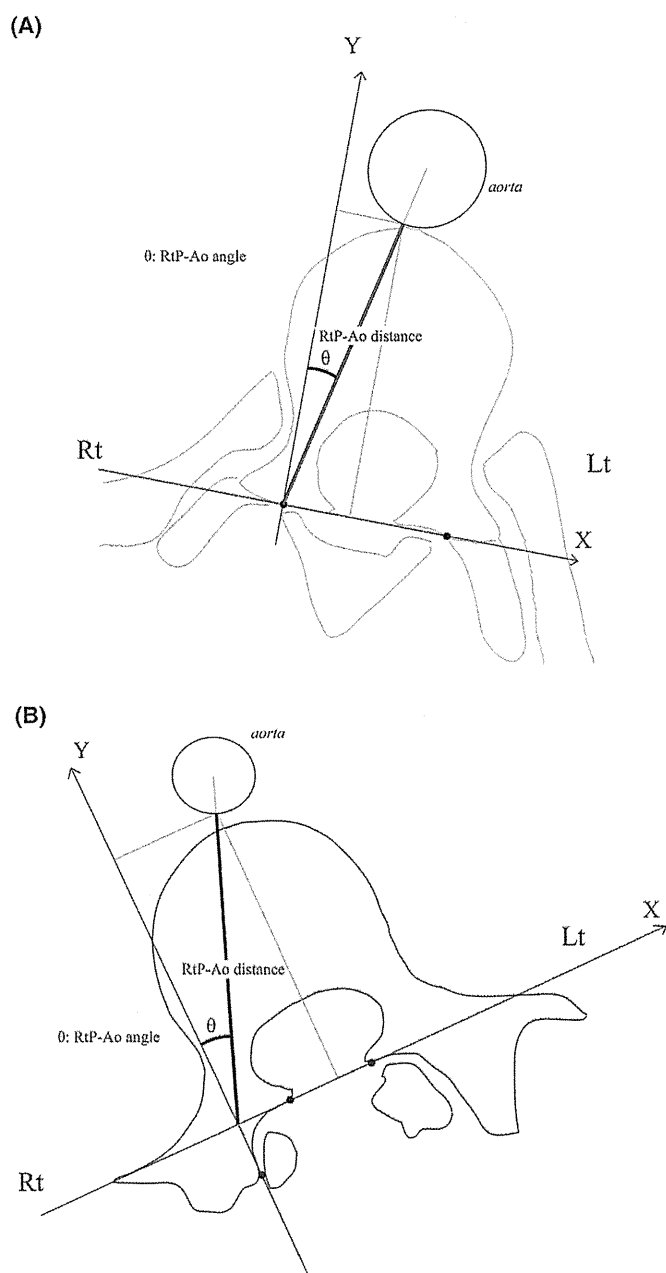


Figure 1. Measurement of two aorta parameters: right pedicle-aorta distance (Rt P-Ao distance) and right pedicle-aorta angle (Rt P-Ao angle). A, Schematic drawing in a thoracic curve. The origin was set at the middle point of the base of a right superior facet, and the X-axis was determined by connecting the middle point of the base of a left superior facet and the origin. B, Schematic drawing in a lumbar curve. The X-axis was determined first by connecting both the medial edges of the superior facets; the Y-axis was drawn perpendicular to the X-axis from the dorsal edge of a left superior facet, and the origin was determined.

trajectory. Sensitivity analysis was performed by variable direction errors and a screw length independently. We defined “aorta-at-risk” when a patient has some level where the simulated pedicle screw involves the aorta. As preliminary analysis had shown that the aorta-at-risk level was observed mostly in the lumbar spine, the maximum error of trajectory was set to

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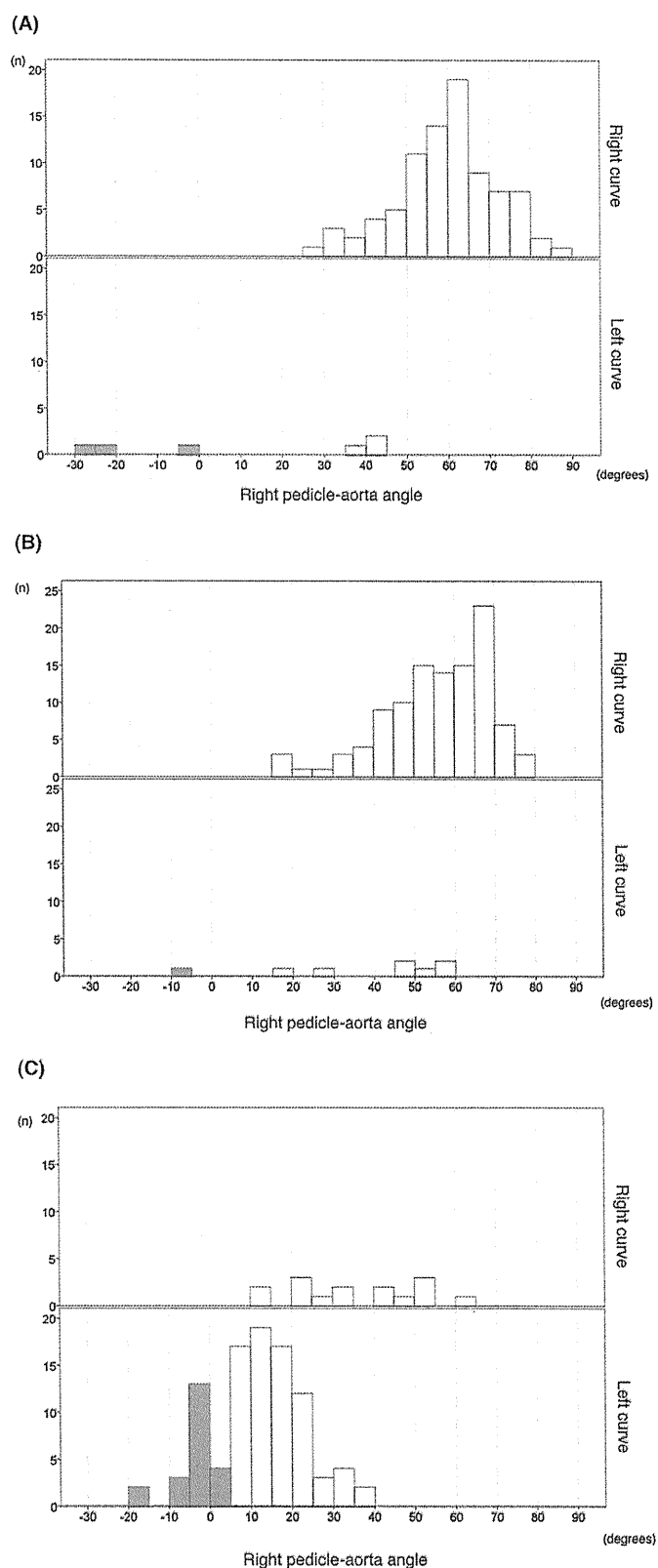


Figure 2. Distribution of right pedicle-aorta angles at 385-spine level. Bar is shaded if a right pedicle-aorta angle was below 5°. A, Histogram at T4 to T7. Upper: right curves, lower: left curves. B, Histogram at T8 to T11. Upper: right curves, lower: left curves. C, Histogram at T12 to L4. Upper: right curves, lower: left curves.

TABLE 2. Distribution of the Aorta-At-Risk Cases With Left Lumbar Curves in Simulated Scenarios

Angle (degree)	Length (mm)	Level	T4 to T11	T12	L1	L2	L3	L4
		Total	147	21	21	21	20	13
5	40	%	0	0	0	0	0	0
		No	0	0	0	0	0	0
5	45	%	0	0	14	5	0	0
		No	0	0	3	1	0	0
5	50	%	0	5	24	29	10	0
		No	0	1	5	6	2	0
10	40	%	0	0	0	0	0	0
		No	0	0	0	0	0	0
10	45	%	0	0	14	10	0	0
		No	0	0	3	2	0	0
10	50	%	0	5	29	33	10	0
		No	0	1	6	7	2	0
20	40	%	0	5	5	0	0	0
		No	0	1	1	0	0	0
20	45	%	0	5	19	10	0	0
		No	0	1	4	2	0	0
20	50	%	0	10	33	33	10	0
		No	0	2	7	7	2	0

The maximum error of medial trajectory of a pedicle screw was 5° in all scenarios.
 Angle indicates the maximum error of lateral trajectory of a pedicle screw; Length, simulated screw length; No, number of the aorta-at-risk cases.

5° in the medial direction and to 5°, 10°, or 20° in the lateral direction, and the length of the screw was set at 40, 45 or 50 mm. We compared the curves between the aorta-at-risk cases and the aorta-no-risk cases. Analyzed curve characteristics were the apical vertebral translation (AVT), the Cobb angle and the Nash-Moe grade.

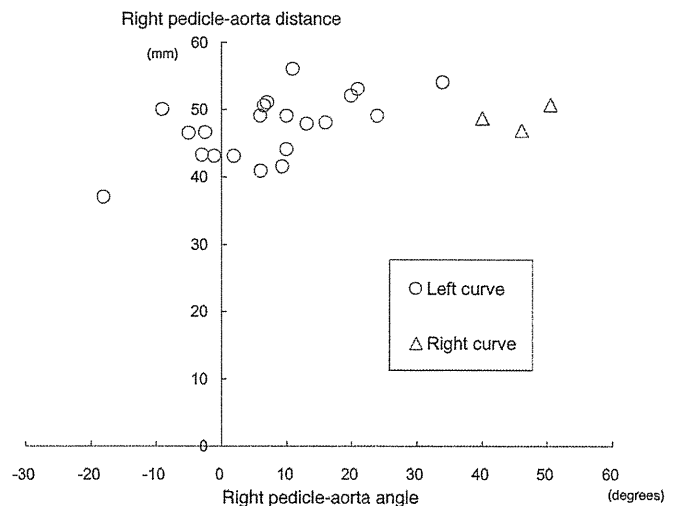


Figure 3. Scatter graph of the aorta location at 1st lumbar level. A circle denotes a case with a left lumbar curve and a triangle denotes a case with a right lumbar curve.

RESULTS

A total of 385 spines were evaluated to locate the aorta position relative to the spine. The aorta often resided in front of a right pedicle at periapical level in left curves (Figure 2). The number of levels where the right pedicle-aorta angle was below 5° was 1 of 2 left thoracic curves and 7 (33%) of 21 left lumbar curves (Table 2). On the contrary, there was no level where the right pedicle-aorta angle was below 5° in 27 right thoracic curves and in three right lumbar curves.

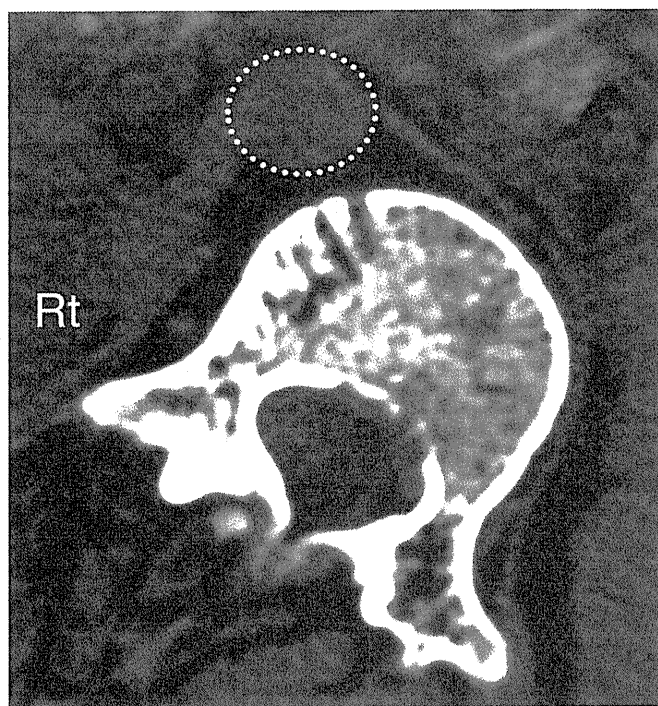
Distribution of the aorta-at-risk cases in nine simulated scenarios indicated that a simulated pedicle screw with a length of 50 mm at L1 or L2 posed potential risk (24%–44%) irrespective of trajectory errors among patients with left lumbar curves (Table 3). Curve characteristics of the aorta-at-risk cases at L1 (Figure 3) were a larger AVT ($P = 0.003$), a larger Cobb angle ($P = 0.006$), and larger Nash-Moe grade ($P = 0.017$) compared with those of the aorta-no-risk cases. AVT, the Cobb angle and Nash-Moe grade were highly correlated with each other: correlative coefficient was 0.814 ($P = 0.000$) between AVT and the Cobb angle, 0.737 ($P = 0.000$) between AVT and Nash-Moe grade, and 0.602 ($P = 0.004$) between the Cobb angle and Nash-Moe grade.

TABLE 3. Past Reports of Aortic Abutment by Thoracic Pedicle Screws Analyzed by Computed Tomography in Patients With Scoliosis

First Author	No. of patients	Follow-up Period (months)	No. of Pedicle Screws	No. of Screw with Aorta Abutment	Left /Right
Liljenqvist ⁶	32	na	110	1 (0.9%)	1/0
Smorgick ⁷	25	na	112	2 (1.8%)	2/0
Sarlak ⁸	19	35.9	185	7 (3.8%)	na*
Total	76		1442	13 (2.5%)	3/0

na indicates not available.
 *Reported with convex/concave. One screw of 7 screws was at the convex side.

(A)



(B)

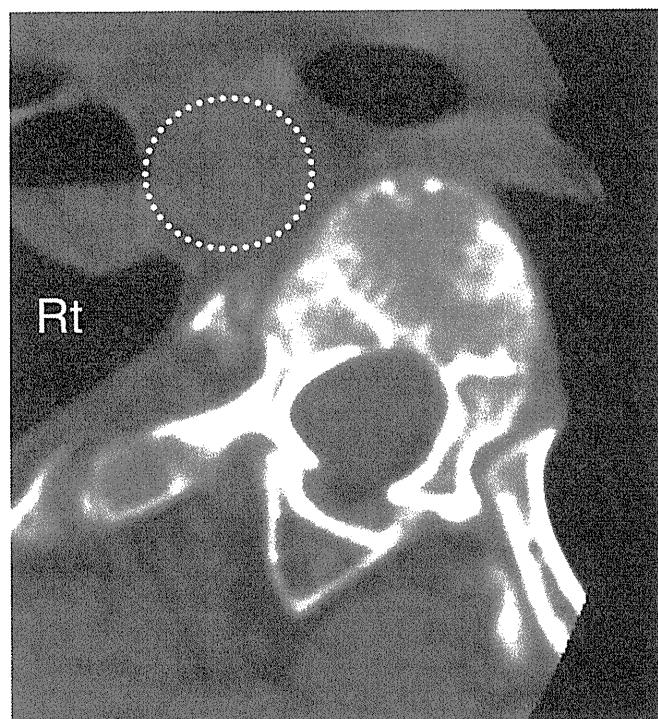


Figure 4. Representative cases in which the aorta resided laterally to the right of the spine. Dotted line circles the aorta. **A,** A 1st lumbar level in a left 88° lumbar curve. **B,** A 6th thoracic level in a left 100° thoracic curve.

DISCUSSION

We investigated lumbar curves in addition to thoracic curves, and found that the aorta moved to the right side of the

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vertebral body in a left curve, and the safety of the aorta from a long pedicle screw decreased either in the thoracic spine or in the lumbar spine. In a case with a large left curve, the aorta shifted to the right lateral side of the vertebral body, and a lateral deviation from an ideal trajectory could result in aorta indentation even by a pedicle screw of a moderate length (Figure 4).

Many spine surgeons prefer to use pedicle screws in thoracic or lumbar spine because of its easiness of placement inside the bony structure and its sound capability in correction and stabilization. However, Kakkos and Shepard⁵ reviewed the delayed aorta injury by pedicle screws in five patients including three deformity cases. They stated, “The true incidence of this complication is probably under-reported” in their conclusion. We found three past studies,^{6–8} which analyzed thoracic curves exclusively by computed-tomography postoperatively (Table 3), and the aorta involvement in 6 (2.5%) of the 1442 screws is substantial. Though the true risk of aortic abutment is unknown, a screw will stay just next to the aorta in such a young generation and it is impractical to monitor such patients closely for over tens of years. In fact, 6 of 76 patients of the past three reports had reoperation.

The aorta resides on the left side of the normal thoracic spine,¹ and past reports dealt with a thoracic pedicle screw on the concave side which is usually the left side of the thoracic curve.^{2,3,7} In scoliosis, a three-dimensional spinal deformity changes the spatial relationship between the aorta and the spine depending on the severity of scoliosis, and the aorta might reside on the right side of a vertebral body. Only one past report by Milbrandt and Sucato⁹ discussed the aorta risk on the right side. They analyzed left thoracic scoliosis and stated “the aorta was in a high-risk position for a (right) posterior screw that exit the lateral border of the pedicle or body in the largest curve of our series.” Our present study confirmed the aorta risk by a right pedicle screw in left thoracic or lumbar scoliosis. We believe a deliberate planning by multiplanar reconstructed images of multidetector-CT is most crucial to prevent an annoying situation. Moreover, we prefer to use a computer-navigation system by preoperative CT for safe placement of pedicle screws.

Judging from high correlations between curve characteristics and the aorta risk in the present study, surgeons need to pay attention to the position of the aorta in placing a pedicle screw on the right in a left curve either in a thoracic or lumbar spine.

Key Points:

- ❑ The aorta is often located in front of right pedicles at the periapical level in left thoracic or lumbar curves.
- ❑ Curve characteristics of the aorta-at-risk cases were a larger apical vertebral translation, a larger Cobb angle, and a larger Nash-Moe grade.
- ❑ Surgeons need to pay attention to the position of the aorta in placing a pedicle screw on the right at the periapical level of a left curve either in thoracic or lumbar spine.

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