CLINICAL DIAGNOSIS OF A PATIENT WITH SPINAL DEFORMITY

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Formulation of expanded diagnosis in spine surgery is of particular importance. This allows for most timely solving the issue of the treatment choice and determining the amount of intervention. A single technique for diagnosis formulation is one of the first steps to establish a national registry of patients with scoliosis, the significance of which needs no justification. The paper presents a methodology for the formulation of a clinical diagnosis in patients with spinal deformities of different etiologies: idiopathic scoliosis, congenital scoliosis and kyphosis, syndromic scoliosis, myopathic deformities of the spine, and Scheuermann's disease. In authors' view, diagnosis should include information on the following characteristics: etiology, side of convexity, localization, structurality and rigidity of the primary curve, degree of deformity compensation, nature of progression and magnitude, sagittal contour of the spine, location of secondary curve, peculiarities of the rib hump, neurological symptoms, pain, age group, comorbidities (vertebral and non-vertebral), and previous surgeries on the spine. In congenital deformities, the abnormalities, both vertebral (intra- and extracanal) and non-vertebral should be noted.
Key Words: clinical diagnosis, deformity of the spine.

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Diagnosis is not a dogma but a guide to action. We remember these words from student years, although at first this idea used to have nothing to do with medicine. A well-known F. Engels in a letter to one F. Sorge (November 29, 1886) criticized German Social Democrats that they treated Marxism in a dogmatic way and did not perceive it as a guide to action. The phase was repeatedly quoted later by the founder of Leninism and it became a winged phrase.

An expanded clinical diagnosis represents in reality a plan of treatment and with the closer and more detailed diagnosis this plan is closer to optimal. The part of vertebrology related to the treatment of patients with spinal deformities of different etiologies is no exception. There is no a single formulation of a diagnosis for these patients in Russia. Frequently, after consultation and examination at different medical institutions the patients come to a specialized clinic and bring many conclusions of healthcare professionals. Any of us have an experience of encountering such diagnoses in these conclusions, which have not been formulated according to standard techniques of diagnosis formulation and often appear rough. "Grade III–IV dysplastic S-shaped thoracolumbar scoliosis" is not the worst option. When one looks more closely, this diagnosis means nothing except for pointing to the deformed spine.

We do not know the papers devoted to formulation of an expanded diagnosis in patients with spinal deformity. A single example of this kind is only given in a monograph by Ya.L. Tsivyan "Scoliotic Disease and its Treatment" [11]: "During formulation of a clinical diagnosis, we believe that it is important not to limit ourselves to mentioning the etiology and intensity of the existing spinal deformity. A clinical diagnosis should reflect the condition of the patient in more detail. Therefore, a diagnosis can be formulated as follows: grade III dysplastic noncompensated right-convex thoracic scoliosis (scoliotic disease) with lumbar counter curvature. Posterior right-sided medial angular rib hump. Spinal functional inability with pain syndrome (radicular, vertebral) complicated with cardiopulmonary deficiency and lower spastic paraparesis".

The authors of this paper are disciples of the Ya.L Tsivyan's school, so there is nothing surprising that the described method is based on the experience of this outstanding surgeon and scientist.

Idiopathic scoliosis

Etiology. Let us consider the most frequent (according to our data, 80 %) spinal deformity. This immediately generates a dilemma. The matter is that following E.A. Abalmasova [1] it is generally accepted to separate a group of dysplastic scolioses based on the presence of dysraphism in a patient. According to E.A. Abalmasova, dysraphism status includes a group of bone dysplasias of the lumbosacral region (spina bifida, sacralization, lumbarization, spondylolysis, spondylolisthesis) as well as neurological symptoms of different grades of intensity

(anisoreflexia, nystagmus, disturbed sensation, vegetotrophic disorders and many others). We have followed these rules for several years and came to the conclusion that this division is irrational and even detrimental. It is irrational because dysplastic and idiopathic scolioses do not differ by any of the following characteristics: disease onset, the course of the disease, orthopaedic status, types of developing deformities, side of the primary curve, prognosis, and choice of treatment. It is detrimental because, according to the worldwide literature, spinal deformities in patients with type I neurofibromatosis belong to dysplastic scolioses [13]. Therefore, we should avoid a terminological bias for the correct understanding of Russian publications worldwide.

Upon detection of any of the above conditions of the lumbosacral region, it should be included in the diagnosis as a separate entity because it can significantly affect the choice of surgical tactics and technique. Neurological microsymptoms do not influence the choice of treatment and should be noted in the records of a neurologist.

The side of convexity. Indication of the convexity side and inclusion of this information in the diagnosis is of a critical significance rather than just a technical step since it is well known that thoracic idiopathic left-convex scolioses more often than right-convex scolioses are accompanied by various intracanal anomalies, mostly syringomyelias. Therefore, many authors perform an MRI study in all patients with left-convex scolioses [19]. We prefer to perform MRI for all patients, and the right choice of this tactics is supported by the fact that syringomyelias are revealed in 12 patients with right-convex scolioses.

The localization of the curve apex (for the primary and secondary curves) is evaluated according to the SRS recommendations:

– thoracic – from the T2 body to T11–T12 disc (the apex of T3–T4–T5 for proximal curves and the apex from the T6 body to the T11–T12 disc for the primary curves);

– thoracolumbar – from the cranial T12 endplate to the caudal L1 vertebral endplate;

– lumbar – from L1–L2 disc to the caudal endplate of the L4 vertebral body.

Structural (nonstructural) scoliosis. A classic paper by Lenke et al. [17] gives a quite clear definition of a structural deformity (both primary and secondary) – a structural curve has a Cobb angle of at least 25° on side-bending radiographs and/or a kyphosis of at least 20° is present at this level, this applies to all localizations of scoliosis. We have not found any other definitions following the publication by Lenke.

Rigidity (mobility) of the deformed spinal region. This section is very important in practical terms because it determines the surgical tactics and technique: whether anterior discectomy is needed, at what length, the necessity of rib head resection? The boundary between rigidity and mobility is not clear, but in our practice, we are guided by the following rule: mobile deformity reduces by at least 30 % at side-bending position.

Compensation. It is measured on radiographs as a displacement value of mid-sacral line relative to the T1 vertebra centroid on anteroposterior spondylograms performed with the patient standing. Scoliosis is rated as decompensated when this displacement exceeds 20 mm. A clinical analogue can probably be a displacement degree of the plumb line dropped from incisura jugularis or spinous process of the C7 vertebra relative to the umbillicus or intergluteal cleft, respectively. It is known that the revealed decompensation in idiopathic scoliosis is rarely noticeable enough to make the patient complain. However, the inclusion of this entity in an expanded diagnosis is reasonable, as it determines the degree of normalization of the shape and position of the spinal column in the postoperative period.

Progression of the deformity. This is one of the most important characteristics of the pathological process in the choice of a treatment method (primarily surgical) and ranking the emergency of the planned intervention. The fact of progression should be confirmed by documented evidences through an analysis of spondylograms performed over a sufficient period (the Cobb angle of the primary curve). The questions immediately arise: what period can be considered sufficient and what dynamics of the Cobb angle indicates reliably the progression of spinal deformity? We have not found definite answers in orthopedic literature. We suppose that an annual (or average annual) increase in the primary curve during 2–3 years by a magnitude exceeding the measurement error in the Cobb method (±3–4°) can be regarded as proven progression of scoliosis. This situation is commonly consistent with the opinion of the patient and his family.

In the literature on the treatment of early scolioses diagnosed before the age of 10 (EOS – early onset scoliosis), the term "malignant progression" is widely used [14]. It is hardly possible to link "malignant progression" to strict quantitative ranges, in addition there are relatively few of these patients. However, it is necessary to include this condition in the diagnosis in all cases, when a surgeon has the impression of rapid unceasing progression, which inevitably leads to early patient disability and incurability resulting in early initiation of urgent treatment.

A 3D-method of examining the posterior topography of the patient's body is an important method helping to look at deformity progression in an objective manner. In Russia, this method was developed in Novosibirsk; it is widely used in screening of school children and is a requited examination procedure during pre- and postoperative care at the clinic of the Novosibirsk Research Institute of Traumatology and Orthopaedics [6].

The magnitude of deformity. The things seem simple – to measure the Cobb angle, correlate with the V.D. Chaklin classification and indicate the measured degree in the diagnosis. This simplicity is deceptive. First, not all doctors who observe patients with scoliosis at the place of residence (more often these are surgeons rather than orthopaedists) can determine the Cobb angle and some use the Ferguson's method, etc. Second, the V.D. Chaklin classification is itself rather

unclear. In 1992, we tried to explore the issue of scoliosis classifications using scoliotic curve magnitude [4]. An analysis of the literature references (the analysis may have been not the deepest) yielded surprising results. Seven (!) of the 15 found classifications were issued from the V.D. Chaklin's pen or were attributed to him by other authors in 1957–1967, with two of them within one year. We continue to use a four-grade classification (up to 10°, 10–25°, 25–50°, >50°), in true, we have to use it since not all patients with scoliosis are girls, and sooner or later boys need to fulfill military service. Military recruitment offices examine recruits for military fitness using this classification.

Another challenge associated with the classification is its discrepancy with modern views on the course of scoliosis. In Russia, deformities up to 10° are classified to grade I scoliosis, while abroad this curvature is a normal variant. Nothing is easy either with grade IV (>50°). Deformities of 55° and 125° belong to grade IV, but any spine specialist understands that they are not only basically different grades of deformity, these are in fact two radically different pathologies requiring appropriate treatment. Therefore, we traditionally include the V.D. Chaklin grading of deformity in the diagnosis, but add this with the Cobb angle magnitude in the brackets for the primary and compensatory curves.

Sagittal contour of the primary curve. The word "scoliosis" is certainly present in the diagnosis, but only in the case if the sagittal contour of the deformed spine is within the normal ranges (thoracic kyphosis is 20–40°). Exceeding these ranges makes doctors pronounce lordoscoliosis or kyphoscoliosis.

The secondary scoliotic curve (counter curvature), by definition, is mentioned the second in the diagnosis after the primary curve. The localization, the magnitude in degrees and structurality are indicated. In the presence of two counter curvatures, the more caudal one, i.e., located closer to the spinal column base, is mentioned first. In cases when the two curves are nearly equal by the Cobb angle, thoracic curve should be regarded as the primary scoliotic curve [17].

Rib hump. The torsional (leading) component of thoracic scoliotic deformity inevitably causes rib hump formation, the type and length of which depend directly on the intensity of the underlying process. The initial and moderate stages of idiopathic scoliosis are characterized by the presence of well-rounded rib hump; in severe deformities there is an angular rib hump. Ya.L. Tsivyan [11] referred to these types of rib hump as lateral and medial, respectively. The presence and type of rib deformity should be included in the diagnosis, because many surgeons believe it is optimal to resect the most deformed portions of the ribs during corrective intervention on the spine, and some perform this resection through a transthoracic access after discectomy [16].

We base on data by E.V. Gubina et al. [2] who showed reduction of a rib hump after correction of scoliotic deformity with partially recurrence within 1.5–2 years. At these periods, following primary surgery, cosmetic resection of the deformed ribs is performed only when the patient expresses a strong desire for this. No more than 10 % of the total number of operated patients expresses this wish. In these cases, we add the diagnosis with the phrase "residual rib hump".

Secondary neurological symptoms. It is well known that the natural course of idiopathic scoliosis, regardless of an age group, is complicated by neurological symptoms occasionally rare. We are not talking about microsymptoms, which can be detected in many patients with idiopathic scoliosis, but about the signs of a conflict which emerged between the walls and contents of the spinal canal. Certainly, this circumstance should be included in the diagnosis and agree with the conclusion of a neurologist.

Pain syndrome. The development of pain syndrome is inevitable because of biomechanics violation of the scoliotic spine and as far as we can judge pain occurs as early as in adolescence. Later pain aggravates and can become persistent to seriously limit the patient's activity. In this case, it is possible to pronounce spinal functional inability. The diagnosis should include pain localization (thoracalgia, lumbalgia, lumboischialgia) and spinal root compromentation.

Age. According to a modern classification, scolioses are classified as infantile, juvenile, adolescent and adult scolioses. Based on the SRS recommendations [7], the first two types are combined in early onset scolioses (EOS). We believe that the EOS group should be mentioned in the diagnosis, because scolioses of the first decade of life are radically different from adolescent and adult scolioses by the disease course and medical tactics, which assumes the most early beginning and multi-staged treatment.

Comorbid vertebral pathology (syringomyelia, Arnold – Chiari malformation, spondylolisthesis, etc.) may require a radical change of treatment strategy with the involvement of neurosurgeons and other specialists.

Previous spinal surgeries. According to the existing rules, a doctor should not use a phrase like "post-operative condition". The full name and date of previous intervention should be provided [3].

Comorbid extra-vertebral pathology is indicated if it may affect the choice of treatment strategy and postoperative course.

The Lenke system [17]. This classification of scoliotic deformities (as well as the rationale for the length of the fusion area) is very widespread in the world. It includes six types of deformities and two modifiers – thoracic and lumbar. As a result, almost any deformity can be described in brief, for example, as follows: 6CN (primary lumbar/thoracolumbar curve, greater than thoracic curve by at least 5°, both curves are structural, upper thoracic counter curvature is nonstructural, the central sacral line is fully located medially to the lateral surface of the apical vertebral body, thoracic kyphosis is within the normal range). Can it replace a detailed diagnosis? It is rather doubtful. Can it supplement a detailed diagnosis? Probably, Yes, especially if an electronic database is used.

A sequence of elements in a clinical diagnosis:

1) orthopaedic diagnosis (the main pathology, comorbid vertebral pathology, comorbid extra-vertebral pathology);

2) neurological diagnosis;

3) comorbid extraskeletal pathology.

An example: grade IV idiopathic right-convex thoracic progressive mobile uncomplicated subcompensated lordoscoliosis (72°) with lumbar structural counter curvature (34°), right-sided well-rounded rib hump, lumbalgia.

Congenital scoliosis

The presented principles of diagnosis formation are applicable to congenital scoliosis, though with one exception. The term "congenital" requires a detailed explanation because impaired development of the spine and thoracic cage, even when systematized in certain groups (segmentations, formations, mixed), is characterized by almost infinite variation. In our opinion, malformations localized in the apex of deformity should be included in the diagnosis only if they are not numerous (1–2), otherwise the diagnosis will be bulky in content and difficult for comprehension. In this situation it is reasonable to speak about multiple malformations of the vertebrae, discs, and ribs. Their detailed description should be given in a protocol of radiographic, CT and MSCT examinations.

In the presence of two equal curves owing to alternating (hemi-) vertebrae, it is necessary to indicate the levels of their localization as the distance between them determines the prognosis of deformity progression.

Deformities with an abnormal vertebra extra the apical zone of scoliotic curve occur rarely. According to A.L. Khanaev [10], these scolioses should be regarded as congenital, as exclusion of an abnormal vertebra from the fusion zone leads to the development of a noticeable frontal imbalance.

Intercanal anomalies (the most common is diastematomyelia) need to be mentioned in the diagnosis; moreover, surgeons are divided as to these abnormalities (either resect or not) [9, 15].

Extra-vertebral anomalies accompany congenital spinal deformities in a significant (up to 76 %) number of cases (cardiac anomalies, maldevelopment of the lungs, diaphragm, gastrointestinal and urinary systems) [8] and should be included in diagnosis, as they can influence the choice of a surgical intervention or be a contraindication to surgery.

Congenital kyphoses

The etiology is clear, but the nature of anomaly requires clarification; moreover, usually this malformation occurs in single cases. Its anatomical features and three dimensional position play a crucial role in deformity progression and complication development.

Configuration of kyphosis (well-rounded, angular) can be important in relation to the pattern of deformity progression.

It is important to take into account the localization of the apex of kyphosis in the development of pathology in the case of affecting the most vulnerable, in terms of blood supply, parts of the spinal cord.

The scoliotic component of deformity can probably be regarded as a favorable prognostic factor, since the presence of lateral inclination of the vertebral canal can provide the dural sac with "the way to sway" to the concave side of the curve to a certain moment and for some time avoid its compression by the deformed frontal wall of the canal.

Neurological symptoms are, in fact, the summary part of the conclusion of a neurologist.

An example: congenital mid-thoracic progressive angular rigid complicated kyphosis (96°) with right scoliotic curve component (23°) due to posterolateral wedge-shaped T6 hemivertebra, compressive-ischemic myelopathy, lower spastic paraparesis with pelvic organ dysfunction, ASIA A.

Syndromic scolioses

A formulation of a diagnosis is the same as in the case of idiopathic scoliosis, except for the etiology of spinal deformity. The main syndrome (type I neurofibromatosis, Ehlers – Danlos syndrome, Marfan syndrome) is mentioned first followed by the detailed characterization of the deformity, which is one of the manifestations of the main syndrome.

Myopathic deformities

In formulation of a diagnosis doctors follow the same principles as with syndromic scoliosis, but a fixed pelvic obliquity should be reflected in diagnosis, as it can be the primary indication for surgical treatment.

Scheuermann's disease

Since the Scheuermann's disease is a mostly single-plane deformity, it is simpler to form a diagnosis than for scoliosis. Note that in contrast to Russia, the world calls this disease as Scheuermann's disease and the name Scheuermann – Mau disease that includes the family name of Dr. Mau is never mentioned. After defining the pathology, we present the stage of development of the pathological process according to S.A. Rheinberg [5]. Afterwards, the localization of hyperkyphosis (usually it is thoracic, rarely thoracolumbar), the magnitude of deformity in degrees and side of scoliotic curve component, if it is present, and finally the localization of pain syndrome are given. In case of the persistent character and poor response of the pain syndrome to conservative treatment that significantly changes the movement pattern of the patient, spinal functional inability should be pronounced as an independent indication for surgical treatment.

It is necessary to remember about the existence of atypical forms of the Scheuermann's disease: changes of vertebral bodies without wedge-shape formation and about thoracic kyphosis exaggeration or kyphosis aggravation without deformity of vertebral bodies [12].

An example: Scheuermann's disease, grade III, thoracic hyperkyphosis (86°) with right scoliotic curve component (14°), thoracalgia.

Complications

Among the complications encountered in vertebral surgery, the most severe, i.e., those leading to re-intervention are as follows:

– neurological due to injury to the dural sac with its contents and spinal cord rootlets;

– purulence – surface and deep, early (up to 30 days after surgery) and late [18];

– implant related complications, i.e., fractures and displacements of endoprothesis elements with injury to adjacent bone structures and without;

– pseudarthrosis of bone block with a significant (over 10°) loss of correction verified before or during revision interference;

– distal junctional kyphosis (DJK) and proximal junctional kyphosis (PJK) should probably be regarded as a complication in cases when they are clinically significant (visible deformity, persistent pain syndrome) and an issue concerning reconstructive surgery should be considered.

All these conditions are included in clinical diagnosis as complications immediately after the name of the performed planned intervention.

An example: grade IV idiopathic right-convex thoracic progressive mobile uncomplicated subcompensated lordoscoliosis (72°) with lumbar structural counter curvature (34°), right-sided well-rounded rib hump, lumbalgia. Correction of spinal deformity with segmental instrumentation, posterior T4–L3 spinal fusion with autobone (May 12, 2014). Fracture of endoprothesis rods.

Conclusions

In surgical vertebrology, a complex field from all points of views, the formulation of an expanded diagnosis is of particular importance. First, it allows one to decide on the treatment choice – conservative or operative. Second, it allows one to determine the amount of a technically difficult and traumatic intervention. A unified technique of diagnosis formulation is one of the first steps to create a national registry of patients with scoliosis, the significance of which needs no justification.

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