doi: http://dx.doi.org/10.11606/issn.1679-9836.v100i4p366-374 Rev Med (São Paulo). 2021 July-Aug;100(4);366-74.

# Disseminated intravascular coagulation: a diagnostic approach

## *Coagulação intravascular disseminada: uma abordagem diagnóstica*

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Andrade AMM, Vieira MS, Horta RD, Chaves MA. Disseminated intravascular coagulation: a diagnostic approach / *Coagulação intravascular disseminada: uma abordagem diagnóstica.* Rev Med (São Paulo). 2021 July-Aug;100(4);366-74.

**ABSTRACT:** The disseminated intravascular coagulation is characterized by a dysregulation in the coagulation homeostasis caused by various conditions. There is a high mortality rate in this syndrome ranging from 45% to 78%, so it is essential to identify. However, it is challenging to recognize, as there are several underlying etiologies, differential diagnoses, difficulty interpreting laboratory tests, and the acute form progresses rapidly. Thus, a narrative review was performed using PUBMED and SciELO to consolidate information for enhanced understanding and accuracy of disseminated intravascular coagulation diagnosis.

**Keywords:** Disseminated intravascular coagulation; Diagnosis; Differential diagnosis.

**RESUMO**: A coagulação intravascular disseminada é caracterizada pela desregulação na homeostase da coagulação e é desencadeada por várias afecções. Essa síndrome possui alta taxa de mortalidade - 45 a 78% - sendo preponderante a sua identificação. Entretanto, o seu reconhecimento é desafiador por existir várias etiologias subjacentes, diagnósticos diferenciais, dificuldade na interpretação de exames e rápida evolução na forma aguda. Assim, foi realizada uma revisão narrativa utilizando PUBMED e SciELO, a fim de estruturar informações para melhor compreensão e acurácia no diagnóstico da coagulação intravascular disseminada.

**Palavras-chave:** Coagulação intravascular disseminada; Diagnóstico; Diagnóstico diferencial.

## INTRODUCTION

The disseminated intravascular coagulation is a secondary syndrome in varied clinical conditions, as sepsis, trauma, pregnancy complications, malignancies, and others affect the homeostasis of the coagulation system. Thus, it promotes its activation and facilitates the formation of thrombi[1,2]. It can result in blood vessel occlusion and decrease the oxygen supply to important tissues and increase the probability of organic failure as a consequence. Concurrently, there is an increased consumption of coagulation proteins and platelets that tend to trigger major hemorrhages and cause death[1,3].

Disseminated intravascular coagulation is extremely clinically important, as it brings about a high mortality rate ranging from 45% to 78%[2]. It is a challenging diagnosis, as it requires greater understanding and correlation to the primary condition of the disturbance4. Besides that, there is still no gold-standard method since the current tests do not independently display the necessary sensitivity and specificity for diagnosing disseminated intravascular coagulation[5,6].

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The laboratory-clinical aspects are essential for basing the diagnosis[4]. Thrombotic thrombocytopenic purpura (TTP), hemolytic-uremic syndrome (HUS), atypical hemolytic-uremic syndrome (aHUS), and coagulopathy in cirrhosis are some of the diagnostic differences in DIC, considering that many laboratory alternations evolve similarly[2,3]. Furthermore, one must abide by the primary causes, such as obstetric complications, traumas, infections, and neoplasms, since they are frequent precursors of DIC[1,7]. Thus, to simplify diagnosis, the *International Society on Thrombosis and Hemostasis* (ISTH) and *Japanese Association for Acute Medicine* (JAAM) scores were created and displayed good results[2].

Therefore, besides score availability, it is necessary to connect them to a clinical rationale for achieving a more effective diagnosis. Thereby, the purpose is to define and understand the main pathologies that can take place in disseminated intravascular coagulation and the primary diagnostic differences to propagate essential information for increased accuracy in diagnosing disseminated intravascular coagulation.

## METHOD

An integrative literature review was performed assessing the Pubmed (The United States National Library of Medicine) and SciELO databanks. English, Portuguese, and French articles were included. They ranged in the period from 1971 to 2019. The indexed keywords are matched to one another as Health Science Descriptors (DeCS) by the "E" Boolean operator, as follows: "Differential diagnosis" And "Disseminated intravascular coagulation"; "Diagnosis" And "Disseminated intravascular coagulation"; "Disseminated intravascular coagulation" And "Causes". In inclusion criteria are: articles that include descriptors in the title or the body of the text, articles that discuss subjects on disseminated intravascular coagulation, their differential diagnoses, the main conditions they trigger, and what the applied diagnostic methods are. Twenty-five articles were selected based on those that related to the purpose of the study for performing the integrative literature review.

**Table 1:** Composite results on the selected x frequency of articles

| | |
|---|---|
| Disseminated Intravascular Coagulation (title) AND Diagnosis (title) | PubMed = 183<br>SciELO = 1<br>Total = 184 |
| Disseminated Intravascular Coagulation (title) AND Differential Diagnosis | PubMed = 137<br>SciELO = 0<br>Total = 137 |
| Disseminated Intravascular Coagulation (title) AND Causation | PubMed = 85<br>SciELO = 0<br>Total = 85 |
| Total | 405 articles |

## RESULTS

405 articles were found based on the descriptors, databases, and predefined period as stated in the methodology. 25 articles were selected from those. That is how a table was prepared explaining the main results found during the critical reading of the articles for preparing the integrative review.

**Table 2:** Articles selected from the bibliographic research.

| Title | Author/Year | Periodical | Main results |
|---|---|---|---|
| How I treat disseminated intravascular coagulation | Levi M, Scully M. 2018. | Blood Am Soc Hematol. | It is a major advance in the early diagnosis of a patient with DIC for improving treatment. Thus, an improved understanding of the predisposing genetic factors of DIC will be helpful to identify the diagnosis. |
| Disseminated intravascular coagulation | Boral BM, et al. 2016. | Am J Clin Pathol | Understanding the physiopathology and differential diagnosis is essential for the treatment of DIC. |
| Pathogenesis and diagnosis of disseminated intravascular coagulation | Levi M. 2018. | Int J Lab Hematol | Description of the definition for DIC, its physiopathology, main causes, diagnostic methodologies, and possible differential diagnoses. |

**Table 2:** Articles selected from the bibliographic research *continuation*

| Title | Author/Year | Periodical | Main results |
|---|---|---|---|
| Current pathological and laboratory considerations in the diagnosis of disseminated intravascular coagulation | Toh CH, et al. 2016. | Ann Lab Med | Early recognition and specific guidance of DIC contributes to a successful treatment. Thus, seeking to identify when the response of the patient changes from protector to maladaptive. |
| Guidance for diagnosis and treatment of DIC from harmonization of the recommendations from three guidelines | Wada H, et al. 2013. | J Thromb Haemost | The utilization of guidelines for leading the diagnosis and treatment of DIC can be effective for putting them into practice. |
| Disseminated intravascular coagulation: What's new? | Levi M. 2005. | Crit Care Clin | Clinical studies have proven that strategies for preventing the activation of coagulation can be beneficial in DIC. |
| Diagnosis and treatment of disseminated intravascular coagulation (DIC) according to four DIC guidelines | Wada H, et a. 2014. | J Intens Care | The DIC categorizations can be bleeding, organ failure, massive bleeding, and asymptomatic cases. Besides that, its treatment and diagnoses must be performed according to four DIC guidelines. |
| Disseminated intravascular coagulation: an update on pathogenesis, diagnosis, and therapeutic strategies | Papageorgiou C, et al. 2018. | Clin Applied Thromb Hemost | DIC is a heterogeneous condition that can be confirmed and requires individualized treatment after analyzing the clinical trials. And for that reason, it requires strict laboratory monitoring. Lastly, there are new treatments that need to be further considered. |
| Disseminated intravascular coagulation: clinical and biological diagnosis | Touaoussa A, et al. 2015. | Ann Biol Clin | DIC's physiopathological mechanisms are complex and depend on the basic pathology, making their clinical expression variable, interfering with the diagnosis, and making consequent treatment faster. |
| Disseminated intravascular coagulation: an update on pathogenesis, diagnosis, and therapeutic strategies | Papageorgiou C, et al. 2018. | Clin Appl Thromb Hemost | Several different disturbances can cause DIC. However, the treatment for the subjacent disease and elimination of the initial mechanism is fundamental. |
| Disseminated intravascular coagulation: a review for the internist | Levi M, Van der Poll T. 2013. | Intern Emerg Med | DIC can cause a generalized condition of fibrin in circulation. Although it is difficult to diagnose, laboratory tests can help, but they are not readily available in diverse clinical surroundings. |
| Advances in the understanding of the pathogenetic pathways of disseminated intravascular coagulation result in more insight in the clinical picture and better management strategies | Levi M,et al. 2001. | Semin Thromb Hemost | DIC causes massive and continued activated coagulation, with consequent bleedings. DIC is a complication from a variety of disturbances, as it is a syndrome. |
| Disseminated intravascular coagulation | Fruchtman S, Aledort LM. 1986. | J Am Coll Cardiol. | DIC is complicating factor of a subjacent disease. It can manifest as abnormalities in a coagulation, hemorrhage, or thrombosis test and cause death. After understanding the physiopathology of DIC and the patient's clinical condition, additional laboratory data, it is possible to perform the diagnosis. |
| Pre-eclampsia | Kahhale S, et al. 2018. | Rev Med. | Eclampsia and HELLP syndrome present an excellent probability of causing complications, such as DIC. |
| Disseminated intravascular coagulation in the HELLP syndrome: how much do we really know? | Haram K,et al. 2017. | J Maternal-Fetal Neonatal Med. | The rate of DIC during pregnancy varies from 0.03 to 0.35%. The leading causes of DIC arising during the gestational period in developing countries are pre-eclampsia and the HELLP syndrome. |

**Table 2:** Articles selected from the bibliographic research *continuation*

| Title | Author/Year | Periodical | Main results |
|---|---|---|---|
| Disseminated intravascular coagulation in obstetric disorders and its acute haematological management | Thachil J, Toh CH. 2009. | Blood Am Soc Hematol. | DIC can occur in situations or scenarios during pregnancy, including emergencies, as premature detachment of the placenta and embolism of the amniotic fluid, and complications from pre-eclampsia. |
| Diagnosis and management of HELLP syndrome | Souza R, et al. 2009. | Rev Méd Minas Gerais | The HELLP syndrome is associated with a tremendous maternal risk of disseminated intravascular coagulation. |
| Differences and similarities between disseminated intravascular coagulation and thrombotic microangiopathy | Wada H, et al. 2018. | Thromb J | Despite the similarity between DIC and thrombotic microangiopathies, there is an activation of all the coagulation, fibrinolysis, and platelet systems in DIC, while in microangiopathies, only the platelets are obviously activated. |
| Thrombotic microangiopathies: thrombotic thrombocytopenic purpura / hemolytic uremic syndrome | Polito MG, Kirsztajn GM. 2010. | Braz J Nephrol | Thrombotic microangiopathies cause diverse clinical and laboratory alterations that can be confused with DIC. Thus, there is a differential diagnosis. |
| Thrombotic microangiopathies: a general approach to diagnosis and management | Arnold DM, et al. 2017. | Can Med Assoc J | Thrombotic microangiopathies are different diagnoses than DIC. The differentiation is important based on clinical and laboratory alterations. |
| Differential diagnosis for sepsis-induced disseminated intravascular coagulation: communication from the SSC of the ISTH | Iba T, et al. 2018. | J Thromb Haemost | It is extremely necessary to recognize DIC early, especially induced by sepsis, to provide adequate handling. Differential diagnoses must be considered and the manner how they must be evaluated. |
| Diagnosis and management of disseminated intravascular coagulation | Karpatkin M. 1971. | Pediatric Clin North Am | DIC can complicate several pathologies, which can result in thrombosis and hemorrhages, even simultaneously. It is even complicated to perform a differential diagnosis on secondary coagulation defects due to the encountered similarities. |
| Current clinical practice DIC 2002: a review of disseminated intravascular coagulation | Toh CH, Dennis M. 2013. | Hematology | Laboratory tests for detecting DIC need to be very accurate and evaluate the risk for improved prognosis of the patient. |
| Diagnosis of overt disseminated intravascular coagulation in critically Ill adults by Sonoclot coagulation analysis | Wan P, et al. 2014. | Int J Hematol | It is challenging to diagnose DIC due to the limitation of the availability of clinical tests or dependable laboratories, as they take longer and are expensive. The study observed patients with identified DIC according to ISTH. |
| Disseminated intravascular coagulation | Gando S, et al. 2016. | Nature Rev Dis Primers | DIC can cause infections (as sepsis) and non-infections (as traumas). The diagnoses are performed based on that; ISTH has established a scoring system on global hemostatic parameters. |

## DISCUSSION

### Disseminated intravascular coagulation

Coagulation physiology is a harmonic process in which all the factors must be balanced to operate adequately[4]. When a blood vessel is lesioned, appropriate amounts of thrombin are produced to form an efficient clot for evolving into a capping mechanism that must be located. There are control measures for that purpose, as the production of anticoagulants to prevent the extension of a clot throughout the entire intravascular system[2,4].

Disseminated intravascular coagulation is a

secondary manifestation to specific diseases caused by an exacerbated stimulation of the pro-coagulant pathways[1,2]. That process is characterized by the excess in the thrombin production due to the stimulation of the tissue factor pathway and the VII factor[1,4]. Thus, there is an exacerbation of the pro-coagulant physiological activities, as the decreased plasmatic levels of anti-thrombin, reduced activation of the C protein, and imbalance of the tissue factor pathway inhibitor (TFPI). Hence, due to the constant stimulation of the hemostatic system, there is an increased propensity for the formation of a systemic thrombosis, as intense hemorrhages[1,2,3].

The clinical and laboratory manifestations present in disseminated intravascular coagulation and its evolution and classification are determined by the underlying disease[8]. Acute disseminated intravascular coagulation can be manifested by hemorrhagic conditions, as gingival hemorrhage, petechiae, epistaxis, hematomas, and gastrointestinal bleeding[9]. Those manifestations are due to the exacerbated consumption of coagulation factors and platelets with the consequent decreased hemostatic capacity[8]. Moreover, due to excess generated thrombin, there is the susceptibility of the occurrence of thrombotic manifestations in the microvasculature and medium caliber vessels that can result from skin lesions, neurological disturbances, gangrene of the extremities, oliguria, and others, that is, malfunction of multiple organs, especially the lungs and kidneys[2,9]. Finally, the laboratory alterations found are: increased fibrin degradation products and do D-dimer, thrombocytopenia (< 50,000/μL) and prolonged prothrombin time (PT), and activated partial thromboplastin time (aPTT)[2].

Since chronic disseminated intravascular coagulation displays continual and non-impulsive activation characteristics that generate a minimum hemostatic malfunction with habitually subtle or nonexistent clinical manifestations, the diagnosis becomes more complex[2,8]. The clinical presentations are predominantly thrombotic, which can be: hyperthermia, hypotension, hemolytic anemia due to microangiopathy, and others. The laboratory alterations can be presented as slight thrombocytopenia and slightly prolonged or normal[2,9].

## Diseases that can be comorbid with disseminated intravascular coagulation

Disseminated intravascular coagulation is a clinical disturbance depending on a subjacent condition for manifestation and determines its acute or chronic evolution[1,3,5,10].

### *Acute disseminated intravascular coagulation*

Severe systemic infections or sepsis are frequent complications from acute disseminated intravascular coagulation. Etiologic agents can be viruses, fungi, protozoans (malaria), especially Gram-Negative or Gram-Positive bacteria[11]. The deregulation process of the coagulation system occurs due to the release of endotoxins and exotoxins from microorganisms that reflects on the inflammation condition from the high degree of released synthesized cytokines by mononuclear and endothelial cells contributing to this process[1,6,12]. The patients who habitually evolve into immunosuppressant conditions are newborns and asplenic. Thus, these patients must be closely monitored who present a condition of bacterial sepsis (Gram-Positive or Gram-Negative) since 30 to 50% of them can evolve into acute disseminated intravascular coagulation[11].

Obstetric complications are other common clinical conditions related to ADIC, and some examples are retained placenta, placenta previa, septic abortion, placenta detachment, amniotic liquid embolism, and eclampsia1[3]. 10% of the patients who suffer from placenta detachment are caused by tissue exacerbated factors in the blood circulation arising from the uterus and placenta, contributing to triggering acute disseminated intravascular coagulation[11].

Since patients with liquid amniotic fluid can retain substances inside of the amnion that will trigger a sudden activation of coagulation and that will bring about an acute disseminated intravascular coagulation condition in over 50% of those patients[12,13].

It is necessary to pay attention to the HELLP syndrome and pre-eclampsia among obstetric complications characterized by arterial hypertension during the pregnancy period and are usually associated with proteinuria and/or edema on the hands and face[14]. Since the HELLP syndrome is characterized by the presence of high levels of hepatic enzymes, hemolysis, and thrombocytopenia, as well as platelet activation, release of inflammatory mediators, and endothelial malfunction that propitiated the disseminated intravascular coagulation condition[15,16,17]. Those two conditions are customarily correlated with pregnancy, considering how the HELLP syndrome can be a complication from pre-eclampsia, and when associated, they are extremely related to developing disseminated intravascular coagulation[14,15].

Severe traumas and burns frequently evolve into acute disseminated intravascular coagulation due to the high release of cytokines in the blood circulation. Besides that, tissue lesions bring on the release of such factors as fat, phospholipids, phosphorus, and thromboplastin that develops in systematic coagulation activation[6,11,12,13]. That process occurs mainly in brain traumas that trigger the high release of thromboplastin and will bring about a condition

of acute disseminated intravascular coagulation[1,13].

Finally, other diseases that can take place in acute disseminated intravascular coagulation are hypovolemic shock, hypoperfusion, acute pancreatitis, hypoxia, hypothermia, hyperthermia, prolonged surgeries, and anaphylactic reactions[1,3,8,10,13].

***Chronic disseminated intravascular coagulation***

Malignancies, such as solid tumors and hematologic neoplasms, can occur in chronic disseminated intravascular coagulation. Their main etiologies occur as breast, pancreas, stomach, lung, prostate, and colon tumors and chemotherapy itself[13]. That process occurs due to the release of tumor cells from pro-coagulant factors, as cysteine proteases for activating the X factor during the cell's life and after its death in chemotherapy[1,6,11,13]. That activity occurs subtly and gradually as it can be asymptomatic and when it shows signs that can be initially identified by platelet deficiency, coagulation factors, and local hemorrhages[3]. There is no measured percentage of cases evolving into chronic disseminated intravascular coagulation in cancer regarding this, but some data confirm the existence of a 20% incidence of metastatic or lymphoproliferative adenocarcinoma[1,3].

Chronic disseminated intravascular coagulation can also complicate vascular diseases. In congenital vascular malformations, the abnormal endothelium becomes covered by tumor vessels releasing an excessive amount of plasminogen activators. Thus, there is an increase of fibrinolysis and fibrinogen and evolves into chronic disseminated intravascular coagulation[3,6,11]. Hence, in situations as gigantic hemangioma and thrombotic microangiopathy, there is the release of the Von Willebrand factor connecting platelets and attract leucocytes and microorganisms to the vessel propitiating chronic disseminated intravascular coagulation[3]. The incidence of this syndrome in patients with vascular diseases is present in 25% of those with massive hemangiomas and only 1% in aortic aneurism[12]. Finally, hepatic diseases also can evolve and end up the same way[1,3,8,10,13].

**Differential diagnoses**

Several differential diagnoses for acute chronic disseminated intravascular coagulation present similar characteristics in its condition, making the diagnosis challenging. There are three cited diseases related to general CDIC diagnosis[18].

A) Thrombotic microangiopathies (TMA) occur due to the formation of platelet clots that cause generalized microvascular occlusions and take place from organ failure, thrombocytopenia, and microangiopathic hemolytic anemia[18,19]. Hemolytic uremic syndrome (HUS), Atypical hemolytic uremic syndrome (aHUS), and Thrombotic thrombocytopenic purpura (TTP) are the primary manifestations of TMA. Meanwhile, obstetric complications as pre-eclampsia are secondary to TMA[18,19,20].

HUS is a bacterial infection caused by the Escherichia coli or Shigella Dysenteriae etiology. Those bacteria release a characteristic toxin named Shiga, responsible for renal microvascular damage (acute renal lesion) and other symptoms[2,19,21]. The most specific clinical symptom is diarreia[2]. The laboratory alterations present in this condition are serious thrombocytopenia, schistocytes, PT, normal aPTT, and normal or slightly raised D-dimer. Besides that, slightly decreased or normal concentrations will be present of ADAMTS13, a protein responsible for the lysis of the Von Willebrand factor[2,3].

(aHUS) is triggered when any stressor stimulus causes a genetic attraction inducing loss of control in activation through an alternative complement pathway. There will be hemolysis, platelet activation, and consequently microvascular lesions as a manifestation[2,21]. The characteristic clinical symptoms of aHUS are vomiting, abdominal pain, bloody diarrhea, pancreatitis, and acute renal insufficiency. The laboratory alterations display normal PT and aPTT and serious thrombocytopenia[2].

Since PTT can be triggered during pregnancy, bone marrow transplant, chemotherapy, and others, and it is caused by decreased activity of ADAMTS[13] facilitates the formation of thrombi and causes microvascular damage and organ ischemia[2,21]. The clinical symptoms of PTT are similar to aHUS, except for the decreased intensity of gastrointestinal and renal symptoms. The laboratory alterations are anemia, schistocytes in the smear, severe thrombocytopenia, ADAMTS[13] activity is severely reduced, and normal PT and aPTT[2,3].

Hence, the similarity of the HUS, aHUS, and PTT with chronic disseminated intravascular coagulation conditions is perceivable, considering that all of them display lesions in the microvasculature, organ failure, severe thrombocytopenia, and reduced ADAMTS[133]. However, in chronic disseminated intravascular coagulation, there is increased PT and aPTT, different from TAMs[2]. Despite this disparity, the diagnosis remains challenging because of variable constants. Due to that, currently, there are professionals in this field who seek to diagnose HUS based on the detection of the Shiga toxin in the organism, and the PTT diagnosis is performed by the decreased emphasis on controlling the Von Willebrand factor and the dosage of ADAMTS[132,21,19].

B) Serious hepatic diseases like cirrhosis and chronic hepatic insufficiency can bring about changes in coagulation, mimicking the findings of acute chronic disseminated intravascular coagulation[2]. Laboratory

changes found in hepatic diseases are: decreased coagulation factors, decreased fibrinogen, increased PT and aPTT, and slight thrombocytopenia[2,3,22]. Thus, the differential diagnosis is complex due to the highly similar laboratory characteristics[3,22]. However, that can differentiate chronic disseminated intravascular coagulation due to the presence of slight thrombocytopenia, slightly increased D-dimer, and the presence of splenomegaly and ascites in the physical exam, which are more suggestive of hepatic disease[3].

C) Heparin-induced thrombocytopenia (HIT) occurs when an antibody links to platelet/heparin factor 4, which forms a multi-molecular complex. Hence, an imbalance occurs in the platelet activation, generating thrombocytopenia and thrombosis. The laboratory alterations are similar to chronic disseminated intravascular coagulation due to the increased PT and aPTT. It is only possible to differentiate them if there is the normalization of these factors after interrupting the use of heparin[2,21].

**Diagnostic Challenges**

It is necessary to analyze clinical and laboratory parameters to diagnose disseminated intravascular coagulation[4]. Global hemostasis tests are used for this, as activated partial thromboplastin time (aPTT), prothrombin time (PT), platelet count, fibrin degradation marker (D-dimer and soluble fibrin monomer), and fibrinogen level[4,9,23]. However, none of these individual tests are conclusive for precisely diagnosing chronic disseminated intravascular coagulation[4,9,24].

The aPTT and the PT are prolonged in chronic disseminated intravascular coagulation. Still, they are not specific, as aPTT is influenced by heparin and by the sensitivity variation of its reagents. PT can be prolonged in other etiologies, as hepatic diseases and vitamin K deficiencies[4,5,9]. There is slight thrombocytopenia in CDIC. However, the same situation can be found in some medications, hemodilution, medullary insufficiency, splenomegaly, or infections[3,4,9]. Since the increase of fibrin degradation, markers, despite being good indicators of chronic disseminated intravascular coagulation, can also be elevated due to some conditions as recent surgery, trauma, or venous thromboembolism. Besides that, if its values are decreased, one cannot ignore possible chronic disseminated intravascular coagulation[5,9]. The high level of fibrinogen and its decrease during the syndrome development are very specific (100%) in chronic disseminated intravascular coagulation, but it displays low sensitivity (22%) as it is a false acute reagent[4]. Besides the peculiarities of each test, it is essential to emphasize that these alterations do not occur synchronously, as a continual repetition is necessary of these exams to obtain the diagnosis. Hence, the experience and insight of the professional are fundamental regarding these data[23,24].

Scores have been developed for this in an attempt to facilitate the diagnostic accuracy of chronic disseminated intravascular coagulation[2]. Currently, there are five scores created by different entities which are: International Society on Thrombosis and Haemostasis (ISTH), the Japanese Ministry of Health, Labor, and Welfare (JMHLW), the Japanese Association for Acute Medicine (JAAM), the British Committee For Standards In Haematology (BCSH) and the Guidelines of the Italian Society for Haemostasis and Thrombosis (SISET) [8,10]. The principles are: The ISTH utilizes the level of thrombocytopenia, the subjacent disturbance, the PT prolongation level, fibrinogen level, and the presence of markers related to increased fibrin as scoring criteria[2,5]. Besides that, the most commonly used score in clinical practice and provides 93% sensitivity and 98% specificity[9,3]. The JMHLW indicates the seriousness of chronic disseminated intravascular coagulation, and it can be used for predicting the results of the disease; The JAAM defined scoring criteria as platelet count, PT, fibrin/FDPs, and specific criteria of the systemic inflammatory response syndrome. That score is more sensitive for identifying patients who suffer from chronic disseminated intravascular coagulation by sepsis, associated with ISTH and JMHLW[5,10].

The scores facilitate the diagnosis process. However, they present failures, as their dependence on laboratory tests must be fast and continual for following up on the development of chronic disseminated intravascular coagulation; although, frequently, laboratories cannot obtain results as quickly[6]. Furthermore, the scores are not efficient for diagnosing asymptomatic chronic disseminated intravascular coagulation[7]. Finally, it is essential to be aware of the subjacent clinical conditions, differential diagnoses, and careful interpretation of alterations in global coagulation tests and hemostatic molecular markers. These factors depend exclusively on the professional and are omitted in them the score criteria[2,7,25].

## CONCLUSION

It is possible to observe the relevance of the chronic disseminated intravascular coagulation diagnosis based on what has been expressed herein, as it is a clinical condition triggered by various diseases and a high death rate. Hence, it is essential to perform a larger number and more accessible studies addressing diagnostic challenges and differential diagnoses. Health professionals, especially recently graduated and inexperienced doctors, know more about this condition to perform diagnoses more quickly and efficiently, corroborating with increased success rates of chronic disseminated intravascular coagulation treatment.

**Participation of the authors:** *Alice Maia Marinho de Andrade*: Research project design and planning; data collection and analysis/interpretation; composition and critical revision. *Mariana Soares Vieira*: Research project design and planning; data collection and analysis/interpretation; composition and critical revision. *Ramona Dias Horta*: Research project design and planning; data collection and analysis/interpretation; composition and critical revision. *Mayra Aline Chaves*: Guidance in the research project design and planning; data collection and analysis/interpretation; composition and critical revision.

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Submitted: 2021, May 03
Accepted: 2021, June 16