SUMMARY
Here we present 4 cases of Knee Sub-Acute Hemophilic Arthropathy according to clinical, X-ray and ultrasound aspects. The X-ray images show subtle changes in the early phases of Hemophilic Arthropathy (HA). Ultrasound aspects are emphasized, which provided direct image of the synovial membrane at the knee anterior capsular recess. We discuss the potential ultrasound use as a marker of inflammatory activity.

Keywords: Ultrasonography; Hemophilia; Knee; Arthropathy; Synovitis

INTRODUCTION
Hemophilia constitutes a group of diseases characterized by a disorder of blood coagulation mechanism, which manifest as a result of an individual’s susceptibility to present spontaneous bleeding episodes or secondary to trauma. Most of hemorrhagic accidents in hemophilic individuals are on locomotive system. The development of factors concentrates has contributed to a longer longevity in this population, leading medical care to be derived to prevention and treatment of orthopaedic sequelae, which constitute the major cause of disability in hemophilia (1,2). The involvement of joints in severe Hemophilia A and B is typical, and called Hemophilic Arthropathy (HA). HA is described as showing three consecutive phases: acute, sub acute and chronic (3). HA’s staging systems are based on X-ray findings(3,4). In these cases, we intended to study the potential of an ultrasound test to study changes on soft parts.

Four cases of HA of the knee are reported here, in which assessment was performed by means of clinical and imaging tests. All patients have been followed up at the Sorocabana Hemotherapy and Hematology Nucleus - Sorocaba Hospital Center - Medical and Biological Sciences Center, Catholic University of São Paulo (NHHS-CHS-CMB-PUCSP). They were all adolescents, males, hemophilic A or B, presenting with a clinical picture of Sub acute HA.

CASES REPORT
Case 1: 12 years old, with a 24-month history of recurrent hemarthrosis on right knee. At clinical examination, he presented with thigh muscle hypotrophy, normal and painless range of motion, joint swelling and palpable thickness of the suprapatelar bursa (figure 1). X-ray images of the knee showed subtle HA signs: osteopenia, epiphyseal enlargement and increased density of soft parts (Figure 2). This was an Arnold & Hilgartner(3) Phase II and the score of Petterson(4) scale was 2. Ultrasound test revealed a joint swelling, synovial thickening and enabled the identification of synovial viliocities (Figure 3).

Case 2: 13 years old, with a 5-month history of recurrent hemarthrosis on left knee. The physical examination was similar to the previous case. X-ray images showed osteopenia, epiphyseal enlargement, increased density of soft parts, angled epiphyseal borders, enlargement of the intercondylar space and some irregularity of the subchondral bone surface. Ultrasound images revealed the same elements as previous case: joint swelling and synovial thickening: 7 mm. This was also an Arnold & Hilgartner(3) Phase II, with Petterson(4) score 3.

Case 3: 15 years old, with a 9-month history of recurrent hemorrhaxis on right knee. At clinical examination, findings were similar to the cases above, with a 10º restraint in extension (Figure 4). X-ray images showed, additionally to the same findings of the previous cases, the presence of subchondral cysts, however not reducing joint space at the femorotibial interline (Figure 5). Ultrasound images showed joint swelling and synovial thickening, reaching 7 mm (Figures 6 and 7). This was an Arnold & Hilgartner(3) Phase III and the Petterson(4) score was 4.

Case 4: 14 years old, with a 24-month history of hemorrhaxis on left knee. At clinical examination, thigh muscle hypotrophy, increased knee volume as a consequence of joint swelling, synovial thickening, and 5º restraint in extension were present. X-ray images showed more significant changes: osteopenia, increased density of soft parts, epiphyseal and intercondylar space enlargement, angularity, subchondral bone irregularity and sclerosis, cysts and reduced femorotibial joint space. This was an Arnold & Hilgartner(3) Phase IV with a Petterson(4)score of 5.

DISCUSSION
The patients were between 12 and 15 years old, and the time of arthropathy development ranged from 5 to 24 months. The older ones (14 and 15 years old) presented with 5º and 10º fexion deformity, respectively. All of them had thigh muscle hypotrophy, joint swelling and palpable synovial thickening.

X-ray studies showed epiphyseal enlargement, increased density of soft parts, and osteopenia in all cases. Subchondral cysts, angled epiphyseal borders, and enlargement of the intercondylar region were evidenced in the three oldest patients; reduction of joint space was seen only in case 4. Ultrasound showed joint swelling in all cases. In case 1, synovial sequelae were all adolescents, males, hemophilic A or B, presenting with a clinical picture of Sub acute HA.
thickening could be seen through the presence of villous dislocations of the posterior salience, as described by Wyld et al.\(^6\) and Hammer et al.\(^6\).

Additionally, we probed the possibility of measuring synovial membrane thickness and found values ranging from 7 to 100 mm, consistent with those found by Bessmeltsev et al.\(^7\): 6 to 8 mm and by Wyld et al.\(^6\): 12 and 19 mm. Those values are higher than normality standards: between 2 and 3 mm\(^7,8,9\). Figure 6 shows an image of joint swelling and of synovial; in Figure 7, the transducer compresses the knee and expels the fluid, providing the synovial thickness measurement. HA in its subacute phase is an arthropathy of inflammatory nature, in which successive hemarthrosis cause synovial inflammation, increasing susceptibility to new hemorrhages, establishing a vicious cycle. The joint suffering from this process during adolescence shall have as a sequel a severe degenerative arthropathy with joint cartilage lesion, fibrous contracture of the joint capsule, and bone injuries. There’s a recommendation towards addressing the synovial membrane as soon as possible in order to avoid damages to joint cartilage and to intervene on natural history of the disease, with the purpose of avoiding joint degeneration\(^2,10\).

When evaluating HA based on X-ray studies, we shall keep in mind that the X-ray tends to underestimate severity of joint changes\(^11\). Indeed, in our study, the X-ray images showed the prevalence of early stages of Arnold and Hilgartner\(^3\)’s HA, with low Petterson\(^4\)’s scores. Nevertheless, the clinical examination was exuberant for all patients, indicating an overt inflammatory activity of the disease. But ultrasound was able to show and differentiate joint swelling from synovial thickening in all studied cases, which were consistent to clinical examination.

CONCLUSION

This study encourages us to investigate the possibility of using ultrasound as a marker of inflammatory activity on knee joint. In case of a treatment such as synovectomy with radioactive colloid\(^10\), ultrasound could be used as treatment outcome control, once it can produce a direct and measurable image of the synovial membrane.

REFERENCES