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## Brief Communication

### The SHOT Project: Study Design

#### Introduction:

Hemophilia has been associated with low bone mineral density (1,2). In the past years several studies have evidenced increased incidence of osteopenia or osteoporosis in patients with hemophilia A and B (3-5): there are many factors that determine a low peak bone mass during adolescence and then, osteoporosis in adulthood. The SHOT (Severe Hemophilia and Osteoporosis Treatment) project will evaluate the muscle-skeletal health status of the patients of the Hemophilia Centre of Reggio Calabria (Italy) by instrumental measurements of the bone densitometry, blood collections and periodic physiotherapeutic evaluations. The purpose is the creation of pharmacological treatment protocols associated to specific programs of physical rehabilitation in the young as well in the elderly.

#### Objective:

The objectives of the SHOT are: evaluate the incidence of osteoporosis in a wide age range of patients with hemophilia, treat pharmacologically those with low bone density with bisphosphonates, coupled with integration of calcium and vitamin D when necessary, and start appropriate rehabilitation programs for prevention of bone loss and fractures for at-risk patients. Other aim will be the improvement of the joint status and the quality of life

of patients with haemophilia and osteoporosis, acting at the same time in terms of pharmacological and physiotherapeutic treatment.

#### Methods:

We enrolled five patients with severe A hemophilia. Age will undergo to six-month hematologic and physiatrie evaluations carried out by expert physiatrist and hematologist. Visits will also include score evaluation based on the joint international scales of measurement, as well as an assessment of quality of life using specific questionnaires. Patients will perform periodic venipunctures in order to investigate the following values: fVIII or fIX activity levels and inhibitor titres, creatinine, total calcium, albumin, phosphorus, alkaline phosphatase, transaminases, bilirubin and prothrombin time, and screening for hepatitis B virus (HBV), HCV and HIV. The examination will be completed by instrumental measurement of bone density.

#### Results:

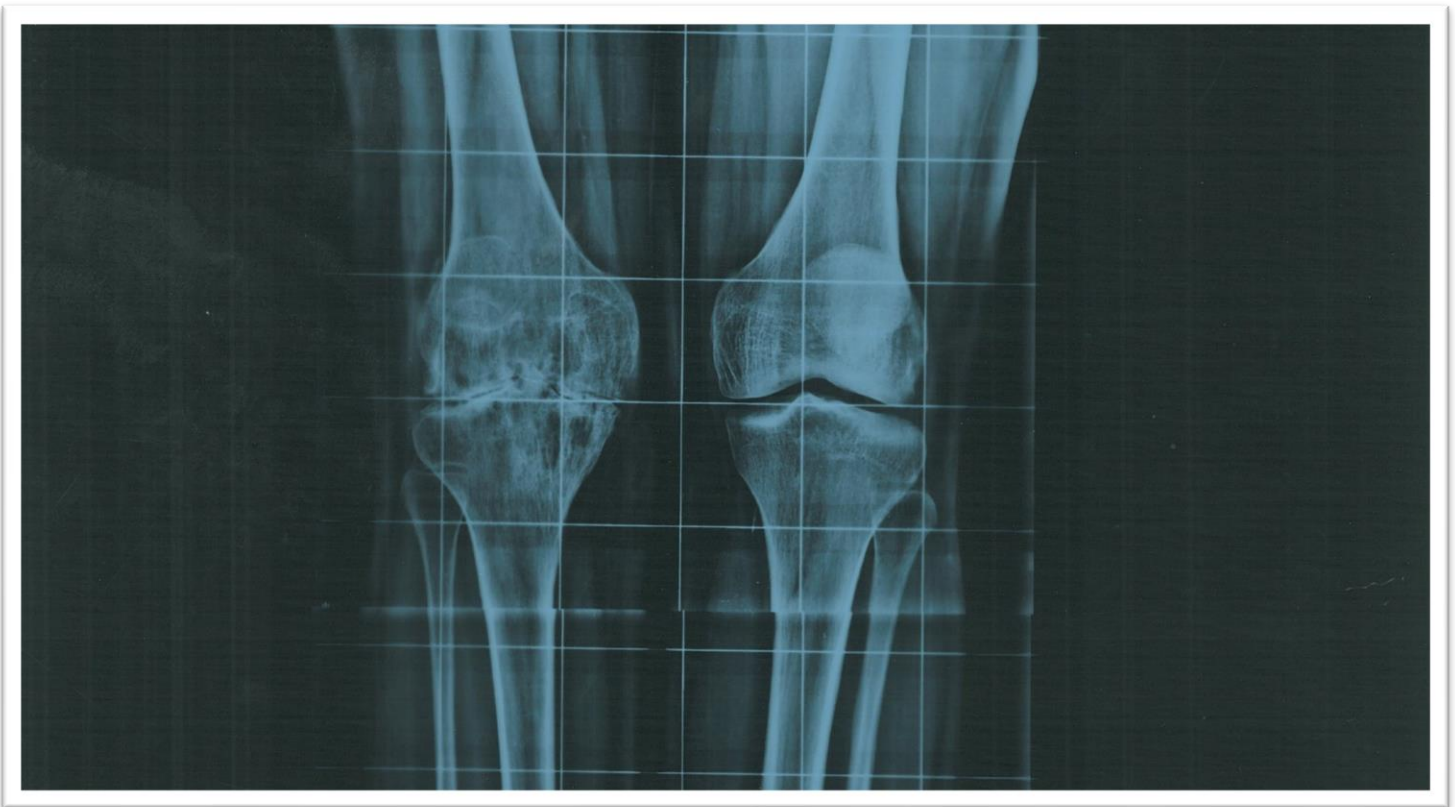
The results of blood tests and instrumental exams (bone mineral density test) after the assessment of the health status, will be used to establish a protocol drug therapy based on



bisphosphonates, calcium and vitamin D and also an individual program of physical rehabilitation tailored to the obtained results as well patients' clinical characteristic. At the end of the project the results will be also analyzed in order to evaluate the statistical significance.

### Discussion:

The expected results from the realization of the SHORT project could be confirm that osteoporosis can be considered as a complication of hemophilia, and also the use of specific combination of drugs of new generation can limit damages from low bone density. This treatment both to an adequate program of physical rehabilitation, may limit the occurrence of arthritic complications, or orthopedic surgery, with resulting in lower costs for the NHS, but also in term of significant improvement in quality of life of patients and their families



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