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Let's talk about

HEMOPHILIA

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HEMOPHILIA

Takeda

Source of information

1. https://www.cdc.gov/ncbddd/hemophilia/facts.html
   - National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention
2. https://www.hemophiliafed.org/understanding-bleeding-disorders/complications/joint-damage/
3. https://www.hemophilia.org/Bleeding-Disorders/History-of-Bleeding-Disorders
4. https://ghr.nlm.nih.gov/condition/hemophilia
5. https://www.wfh.org/en/resources-education/educational-materials
6. http://www1.wfh.org/publications/files/pdf-1586.pdf
7. https://www.hemophilia.ca/files/Home%20Treatment%20Guide.pdf

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What is hemophilia?[1]

Hemophilia is a rare condition that affects the blood's ability to clot. It's usually inherited, and most people who have it are male. People with hemophilia have the deficiency of clotting factor, which results in delayed or deficient clot formation after injury. This means they bleed for longer than usual.

What are the different types of hemophilia?[4]

Human body depend on many types of proteins produced in the body to initiate blood clotting and to stop bleeding. Type of hemophila is based on the type of protein that the body either does not produce enough or is missing in the body.

The most common types are hemophilia A and hemophilia B.

A — Hemophilia A (Classic Hemophilia) caused by a lack or decrease of clotting factor VIII in blood.

B — Hemophilia B (Christmas Disease) caused by a lack or decrease of clotting factor IX in blood.

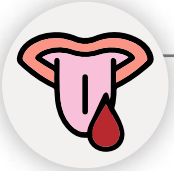
How do we know that the child is a hemophilia patient?

Hemophilia is suspected and confirmed when babies have[1,4] :

Prolonged bleeding or oozing following an injury, surgery, or having a tooth pulled

Spontaneous bleeding episodes (bleeding without any major injury) that most often occur in muscles, joints, and soft tissue

Blood in the urine or stool



Bleed longer than usual after an injury, especially to the mouth and tongue.

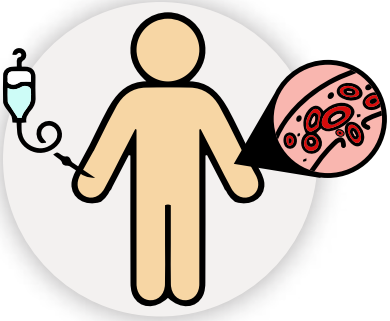
What tests do doctors advise to confirm hemophilia?[1]

Many people who have or have had family members with hemophilia, need to check their baby boys after birth.

Doctors may recommend screening tests and/or clotting factor tests.

Screening tests are blood tests that show if the blood is clotting properly. Clotting factor tests, also called factor assays, are required to diagnose the type of hemophilia and the severity.

How is hemophilia treated?[1]



The best way to treat hemophilia is to replace the missing blood clotting factor (popularly called factor) so that the blood can clot properly. This is done by infusing (administering through a vein) commercially prepared factor concentrates.

Doctors typically prescribe factors for

1 **Episodic care (On-demand therapy)** to stop a patient's bleeding episodes

2 **Prophylactic care (Preventive therapy)** to prevent bleeding episodes from occurring

Since each person responds to treatment differently for a variety of reasons, an analysis of specific symptoms—such as bleeding rates and level of factor in the blood—can help doctors find the right treatment and regimen for a particular person with a bleeding disorder

What is On-demand therapy?[3]

On-demand therapy is factor replacement therapy given to stop uncontrolled bleeding after bruising or injury. Normally patient is taken to doctor or hospital where he is advised right dose and administered the infusion.

What is prophylaxis or prophylactic treatment?[6]

Prophylaxis is the regular use of clotting factor which prevents or reduces the incidence of bleeding episodes. Injections of clotting factor are given one, two or three times a week to maintain a constant level of these factors in the bloodstream so that severe bleeding episodes are avoided.

Use of adequate clotting factors is becoming the normal mode of treatment for younger patients, and can be started when the veins are well developed (usually between the ages of two and four years).

How do repeated bleeding episodes affect the body?[5]

In hemophilia patients, slightest injuries may lead to bleeding in joints. Repeated bleeding into a joint causes the synovium (lining of the joint) to swell and bleed very easily.

Some blood remains in the joint after each bleed. The synovium stops producing the slippery, oily fluid that helps the joint move.

This damages the smooth cartilage that covers the ends of the bones. The joint becomes stiff, painful to move, and unstable. It becomes more unstable as muscles around the joint weaken.

With time, most of the cartilage breaks down and some bone wears away. Sometimes the joint cannot move at all. The whole process may lead to permanent damage called hemophilic arthritis. This leads to disability.

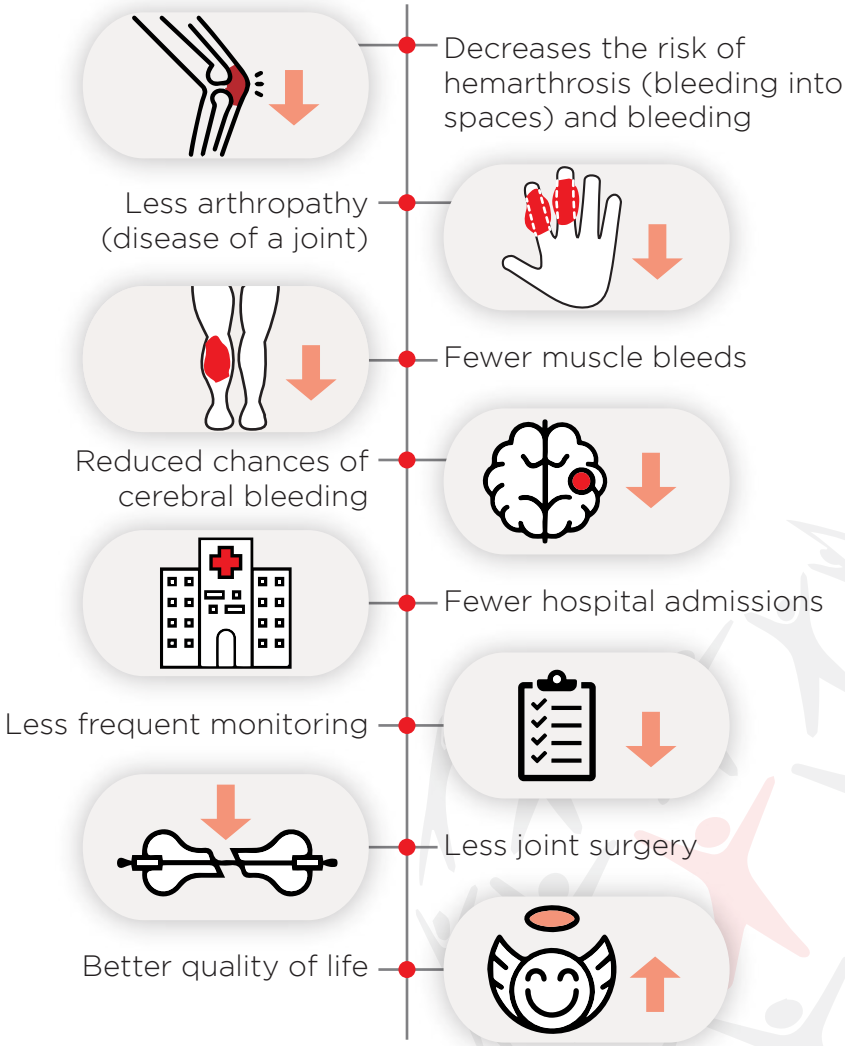
Why prophylaxis for your child?[1]

Prophylaxis is a preventive measure in hemophilia where regular infusion of blood clotting factor concentrates helps avoid bleeding.

Why compliance is important?[5]

Prophylaxis can help reduce or prevent joint damage and improve the quality of life of people with hemophilia. However, these outcomes are only achieved with adherence to prescribed prophylaxis regimens i.e., regular infusion as suggested by your doctor.

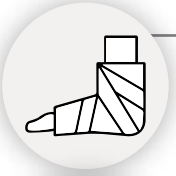
Advantages of Prophylaxis[1,5]



What happens if you don't give prophylaxis?[3]

- Bleeding episodes - joints, muscles, soft tissues
- Progressive joint damage
- Time lost from school or work
- Reduced activity level when bleeding present
- Physiological and emotional concerns
- Insurance issues

Do's and Don'ts when on Prophylaxis[5]

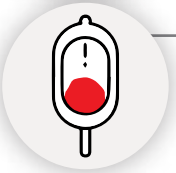


Rush immediately to nearest centre if got hurt with no swelling but unstable or unsure; or got hurt and unable to walk.

During exams take small breaks while writing. This is to avoid pressure on shoulder, elbow and wrist while writing for 2-3 hours.

Get a letter from your centre/doctor to allow your child 20-30 extra minutes for each exam or arrange writer.

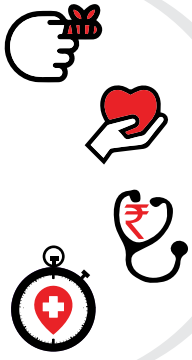
Do not ignore continuous headache as migraine. This could be due to bleeding in brain (brain hemorrhage).



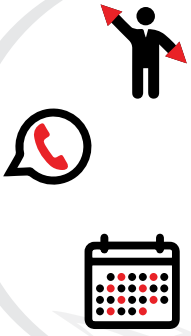
Do not ignore abdominal pain or blood in urine. Consult your doctor immediately.

Home infusion tips and care:[7]

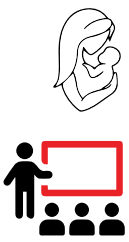
Home infusion makes it possible to give prompt treatment at the first sign of bleeding and makes it easier for families to follow prophylaxis therapy.



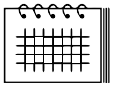
- allows the family to treat bleeding at the earliest possible moment. This is the best way to reduce damage to joints and to speed recovery.
- helps the family gain more confidence in caring for the child with hemophilia and a greater feeling of control over their lives. It also allows them to travel more easily.
- decreases overall health care costs because fewer visits are made to the hospital.
- avoids the delays in travelling to the hospital, assessing the child there and prescribing treatment.
- at the time of hospital visit learn to give factor under the guidance of hospital staff
- ask them to send message or WhatsApp every time they give the factor at home and main a register to note each dose with dates.

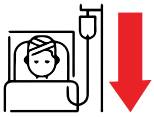


- makes it easier for the child to follow a prophylactic treatment program, designed to prevent joint disease from occurring in children. Most prophylaxis programs require treatment on a regular schedule of 2 or 3 times per week. This can be hard if the family is required to travel to hospital for each infusion.



- Children are sensitive. To avoid pain to them while giving factor, dummies can be used while training parents





- once trained, allow them to give factors at home but give only one month dose and should mandatorily visit centre every month. Ask them to bring all empty vials and monitor doses.
- reduces the number of in-hospital days and outpatient visits. This means that the child misses less time from school and the parents miss less time from work and family.
- helps the child and family to be more independent of the health care system.
- reduces disruption to the family's daily activities.
- allows the child to participate in normal activities, including, most sports.
- fosters a strong partnership between the family and the comprehensive care team.

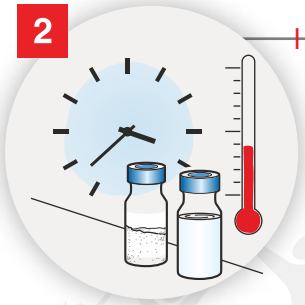
Infusion Instructions for Factors[3]

(Depending upon the product package insert guidelines for infusion)

1

Wash your hands with soap and water thoroughly

2



Take the contents of the pack out from fridge; leave standing to bring to room temperature

3

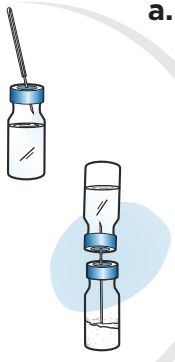
Get the Infusion kit/ tray ready

a. 2 surgical sprint/disinfection swabs for disinfection of the vials
b. Sticky tape to keep the butterfly needle still, plaster and cotton wool
c. Alcohol swabs for skin disinfection
d. Syringe and tourniquet
e. Bottle of factor (powder) and diluents (liquid)

4

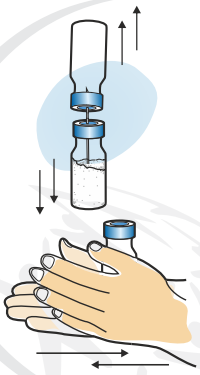
Apply local anesthetic to the site of the vein

5 Getting the factor ready for infusion (reconstitution)



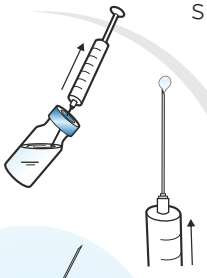
a. Remove protecting covering from short end of double-ended needle and insert exposed needle through stopper to the diluents vial

b. Remove protective covering from long end of double-ended needle. Invert solvent {diluents) vial over the upright factor (powder) vials, then rapidly insert free end of the needle through the factor (powder) vial stopper at its center. The vacuum in the vial will draw in the solvent



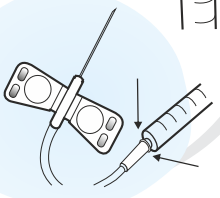
c. Disconnect the two vials by removing needle from solvent vials stopper, then remove needle from factor (powder) vial

d. Roll the vial gently between the hands **without shaking it,** until the solution is clear. Vigorous shaking to be avoided



e. With the filter needle on the syringe; draw in the reconstituted factor into the syringe

f. Expel air out of the filter needle {if any)

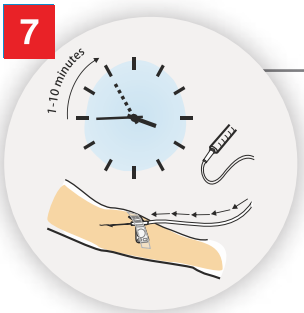


g. Discard the needle into the sharp box, and connect the syringe to the tubing of butterfly needle

6

Put on the tourniquet, disinfect the injection site with the alcohol swab and insert the butterfly needle with bevel facing upwards and secure it with adhesive tape. Check that the needle is in the vein by drawing little blood in to the butterfly needle, and undo the tourniquet and inject the fluid, checking the back flow from time to time

7



Inject slowly; the injection should last for 1-10 minutes

8

Remove the needle and put a dry swab on the injection site

9

Put on last adhesive bandage and clear up everything

10

Note down the date of injection and the reason why

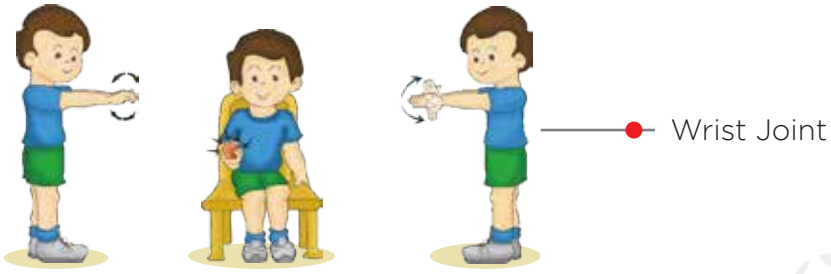
Home exercises/Physiotherapy:[2]

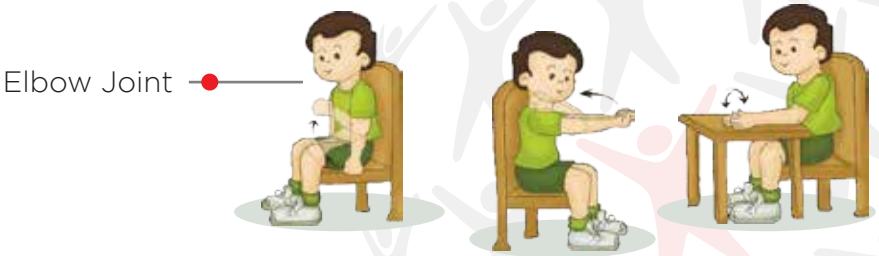
Physical activity should be encouraged to promote physical fitness and normal neuromuscular development, with attention paid to muscle strengthening, coordination, general fitness, physical functioning, healthy body weight, and self-esteem.

Kids with hemophilia can participate in activities, though they might have to take on a different role. For example, hemophilia might prevent kids from participating in contact sports, but they can still be a part of the team as the scorekeeper or assistant manager. Non-contact sports such as swimming, walking, golf, badminton, archery, cycling, rowing, sailing, and table tennis should be encouraged. However, care must be taken when involving in any sport or physical activity.

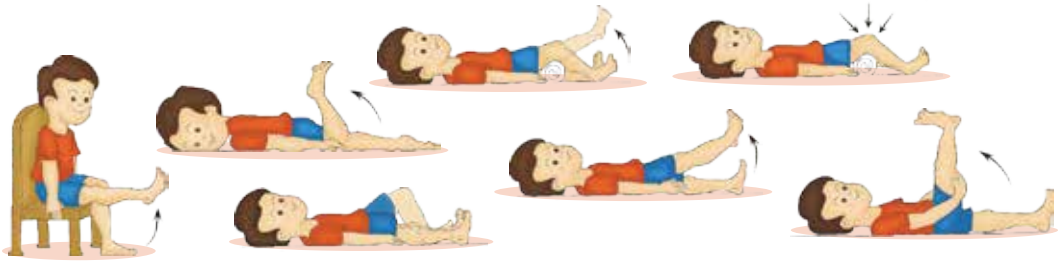
A card should be provided to capture physiotherapy sessions. Physiotherapy once a month and continue same exercises twice a week.

Physical Exercises in Hemophilia:





Knee Joint



Hip Joint

Shoulder Joint

Ankle Joint

School Tips:[3]

By the time they reach school age, children are generally capable of thinking logically and seeing cause and effect. A school-aged child will be able to report when he has a bleed. As children get older, it's helpful to involve them as much as possible in their own care. This can include teaching them on how to track their injuries. But if you react to bleeding episodes with anger, fear and frustration, your child may try to protect you by hiding a bleed even when his pain is hard to bear.

Following are few tips for schooling of your hemophilia child.

Allow kids to attend school regularly

Always wear knee pads

Inform teachers and school authorities

Guide them with do's and don'ts for school (injuries to be informed immediately)

Tell them about using R.I.C.E. (Rest, Ice, Compression, Elevation) for bleeds

Factor infusion to help stop the bleed

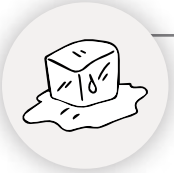
Provide emergency contact details

Maintain frequent communication with school teachers

Ensure factor taking on-time and how parents must deal with kids[3]

It is important to take your factors on time to avoid the risk of bleeding.

Maintain an accurate up-to-date diary of all infusions given, which includes the bleeding sites, the amount of factor and other first aid measures used.



Along with clotting factor, use ice, compression and elevation of limbs to help reduce the bleeding.

Maintain frequent communication with your doctor.

Know what to do for allergic reactions, and report them promptly.

Educate and encourage your child to be aware of his body so that he can recognize and report bleeds, Involve your child in his own health care.

Store and dispose of home infusion supplies (needles, syringes, etc.) safely.

Store clotting factor concentrates according to the instructions provided.

Precautions during vaccination and dental procedures[3]

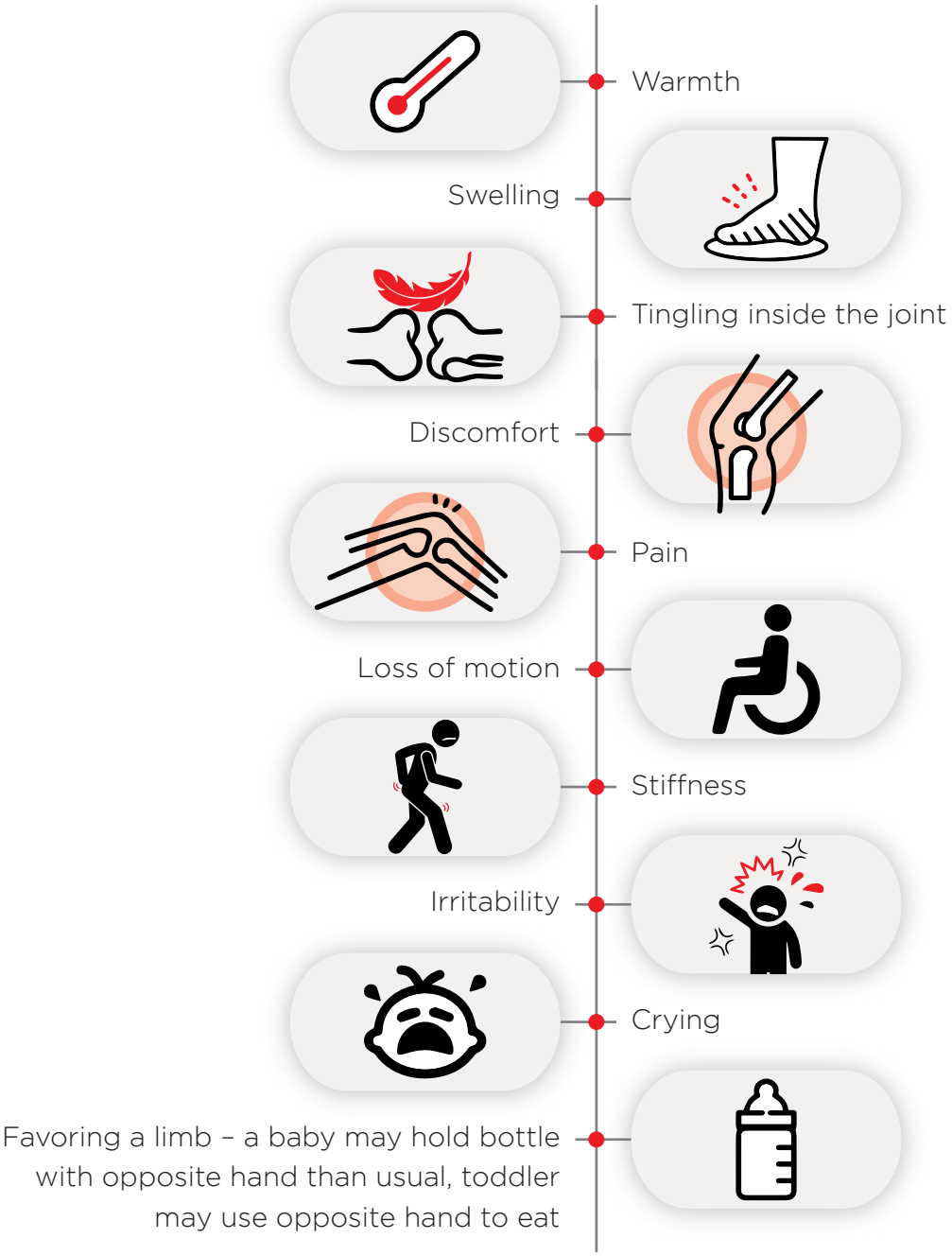
To help prevent bleeding problems, doctors use caution when treating children with hemophilia. For example, when giving immunization shots that are normally given in the muscle, doctors instead inject the shots into a deep area under the skin called the subcutaneous tissue. It's important to remember that kids with hemophilia need all recommended vaccines.

Make sure that your child is brushing his or her teeth twice a day, flossing regularly, and drinking fluoridated water to keep the teeth as healthy as possible. This will make it less likely for the gums to bleed or for your child to need dental surgery. Routine cleanings can sometimes cause bleeding. Find a dentist who has experience with patients who have hemophilia and who knows how to handle bleeding if it happens.

Don't ignore internal bleeding. Keep joints safe.[3]

Bleeds can happen in or near joints such as the ankles, knees, hips, elbows, and shoulders. It is important to treat these bleeds as quickly as possible to stop the pain and bleeding and prevent a more serious bleed that is difficult to control. The more blood in the joint, the more damage it can cause.

Observe the symptoms of a joint bleed[2]



Prophylaxis prevents bleeding and joint destruction and should be the goal of therapy to preserve normal musculoskeletal function.[5]

STOP IRREVERSIBLE JOINT DAMAGE. START PROPHYLAXIS.

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