Measuring the blood pressure in patients with shortened arms due to thalidomide embryopathy

Background:

Pathologically elevated blood pressure is a major risk factor for developing severe complications in terms of heart disease and arteriosclerosis (hardening of the arteries). Untreated high blood pressure leads to long-term multi-organ failure (heart, brain, kidneys, eyes).

This and the fact that high blood pressure usually goes unnoticed by the patient (silent killer), measuring the blood pressure on a regular base is one of the most important actions that can be taken to prevent severe future organ failures.

As technical device for this diagnostic approach the method of Riva-Rocci, modernized by Korotkov and Cushing has been successfully used for more than a century:

The cuff is placed snugly around the upper arm, at roughly the same vertical height as the heart while the subject is seated with the arm supported. Localisation of the cuff and correct cuff diameter are essential for an exact and correct measurement of the blood pressure. Too small a cuff results in too high a pressure, while too large a cuff results in too low a pressure.

A variety of cuffs with different diameter are accessible. Bigger arms require larger diameters of the cuff.

While listening with a stethoscope to the brachial artery at the inner side of the elbow, where the brachial artery is located, the pressure in the cuff is inflated to a pressure of approximately 200 mm mercury, a pressure where complete occlusion of the arm artery can usually be expected and the examiner does not hear any sound through the stethoscope. The pressure in the cuff is then slowly released via a valve. As the pressure in the cuffs falls, a "whooshing" sound is heard as soon as the blood flow first restarts in the artery. The pressure at which this sound is perceived is recorded as the systolic blood pressure. The cuff pressure is further released until the sound can no longer be heard (the artery not being compressed any more). This is recorded as the diastolic blood pressure.
Digital instruments (which are not as exact as the “old fashioned” manual mercury devices and which need regular calibrating) use a cuff, which may be placed around the wrist. It is paramount that the sensors of these digital instruments are at the exact site of the radial artery.

The blood measurement via the Riva-Rocci method, which exists since more than a century is a very reliable, exact, easily accessible and easily applicable, cheap and non-invasive method. In terms of exactness, it is only surpassed by an intra-arterial blood pressure monitoring system which is invasive, expensive, time-consuming, prone to complications and therefor restricted to use in intensive care monitoring and during surgery.

As mentioned above, a correct measurement strictly depends on the following factors:

1.) Exact blood pressure measuring device
2.) Correct diameter of cuff depending on arm diameter
3.) Cuff must be snugly fixed in the height of the heart
4.) Hearing of doctor is unimpaired

5.) Upper arm with normal anatomy must be patent
6.) Diameter of arm artery must be in normal relation to diameter of upper arm

Factors 5 and 6 are often not patent in thalidomiders with upper extremity deformation.

If the "upper" arm is patent at all, it is often of a non-consistent and more conic diameter, which leads to a slipping off of the cuff. Aberrant or too small arteries of the arm will most certainly lead to a situation, where the "whooshing" sound is not heard during auscultation with the stethoscope or – even worse – at a wrong pressure of the inflated cuff. Small in diameter arteries will be compressed at a lower cuff pressure as normal arteries would and will lead to the assumption of a normal blood pressure while the blood pressure is elevated in reality.

Patients with normal arms have difficulties to manage correct application of the cuff at the upper arm. This is, why the cuff is applied by a helping second person in doctors offices and for this reason, wrist cuff devices were developed to foster self-management of daily blood pressure monitoring.

A wide variety of wrist cuff devices for blood pressure measurement of different quality are available on the market and the german "Hochdruckliga" ("High pressure liga") (http://www.hochdruckliga.de) publishes new certified devices which yield correct data in patients with normal wrists. (http://www.hochdruckliga.de/messgeraete-mit-pruefsiegel.html),

As mentioned above, wrist measurement devices require a normal anatomy of the wrist to yield reliable results. Because of there altered anatomy of their arms, thalidomiders with upper extremity defects will not be able to use upper arm cuff devices because the devices cannot be handled alone and because the blood pressure measurement results will be false. Wrist devices may be handled with short arms but will yield wrong results due to aberrant or missing radial artery.

Hypertension and short arms:

Blood pressure monitoring in persons with short arms may be even more important than in other patients because investigations[1] show that arterial hypertension may be more frequent in patients with posttraumatic limb amputations than in patients with normal extremities.
The reason for this remains to be elucidated, reduced overall diameter of blood vessels may be one of the reasons. There are no studies about an exact pathomechanism or about the question if this observation was also made in congenital limb defects as the thalidomide embryopathy.

<u>Blood pressure measurements in thalidomiders with upper extremity damage: Problems:</u>

Management of device:
Even with a wrist cuff device, this can often not be managed with short arms. Help by a partner or a health care provider could be a feasible solution.

[1] Systolic arterial hypertension in patients amputated for injury. Labouret G, Achimastos A, Benetos A, Safar M, Housset E. Presse Med. 1983 May 14;12(21):1349-50

Incorrect blood pressure results:
Reliable wrist cuff devices which are available since approximately 10 years yield precise results in patients with normal wrists, excellent devices demonstrating the correct position of the pressure sensor[2].
Various devices with different cuff diameter are available, meeting the different diameters of human wrists.

- These wrist cuff devices are known to often produce error messages when used by thalidomiders with deformed arms and in case of "correct" pressure results, these seem not to be reproducible, implying that the results are far from correct.

- Upper arm cuff devices rarely work at all in thalidomiders with arm damage. Apparently, aberrant anatomical position or diameter of the radial or brachial artery seem to yield unreliable blood pressure test results in this patient group.

- There are Thalidomide cases in which only very slight upper extremity damage like hypoplasia of thenar bulge is apparent but MRI findings show complete missing of the radial artery. This shows that extensive vascular malformation may exist in spite of very little deformation of upper extremity. Wrist cuff devices will yield no or wrong data in these patients.

- Arm diameter changes in a very short distance from shoulder to "elbow" in patients with upper extremity deformation due to thalidomide leading to a "conical" arm profile which results in "slipping down" of the upper arm cuff blood pressure devices. (see picture)

- It must be assumed, that the ratio of the diameter of the blood vessels / diameter of the arm in thalidomiders is different from persons with normal arms due to reduced amount of tissue which needs blood supply. Duplex ultrasound of thalidomiders showed reduced diameter of arm vessels even in normal upper arm diameters[3].

[2] http://www.aerzteblatt.de/archiv/38610/Diagnostik-Automatische-Positionierung-fuer-Handgelenk-Blutdruckmessung?s=blutdruckmessen
[3] personal observation in different patient data from assessments fort he thalidomide trust

- Eye investigations in thalidomiders with upper arm defects have shown severe signs of long standing arterial hypertension – in spite of the family doctor always having measured "correct" blood pressure values.

This implies, using regular blood pressure devices in thalidomiders will risk to show no or false results.

This patient population being 50 years now, finding reliable blood pressure measuring methods seems mandatory.

Invasive intravascular arterial blood pressure measurement:
This investigation is usually limited to ICU treatment or surgical therapy but may be useful for the individual in case surgery is planned anyway.
If non invasive external blood pressure measurement on arms or legs yields to reproducible results at all, simultaneous intraarterial measurement during surgery may provide correct data that can be used to "interpolate" external pressure measurements with a "correction factor" based on this simultaneous measurement.

Blood pressure measurement on lower extremities:
This is used as a standard in situations where blood pressure measurement on the upper extremities is not possible and yields blood pressure of 20% above the correct value. Usually assessment of only the systolic value is feasible, good enough for shock and trauma management but insufficient for regular measurements to prevent cardiovascular disease from arterial hypertension, where the diastolic ("lower") value of the blood pressure measurement is the important factor.
On top of this, arteriosclerotic lesions (which are more frequent on lower extremities than on arm vessels) will falsify the results of a conventional blood pressure measurement:

- Diabetic mediasclerosis leads to rigid blood vessels lead to enormous resistance against pressure from outside and will yield false high blood pressure levels in measurement of blood pressure.

- Arteriosclerotic lesions as in "claudicatio intermittens" show reduced blood vessel diameter due to arteriosclerotic plaques leading to rapid occlusion of arteries by external pressure and yielding false low blood pressures in blood pressure measurements.

There is very little data available to blood pressure measurements in thalidomiders.

Shiga e.a. [4] have found an excellent accordance between diastolic blood pressure measured with either invasive (intrarterial) and noninvasive devices (the latter beeing cuffs from Nihon Kohden and Terumo), applied above the arteria tibialis posterior. The systolic blood pressure was 10-20 mm hg higher in the non invasive measurements than in the invasive techniques. Yoshizawa[5] comes basicly tot he same results and reccommends the formula ((systolic

[4] Int J Surg Case Rep. 2014;5(7):428-30. doi: 10.1016/j.ijscr.2014.05.002. Epub 2014 May 20. Shiga Y, Nojiri F, Yoshizawa A, Shimbo T, Kawachi S. Measurement of blood pressure in a thalidomide-impaired patient who required ovarian cystectomy: A case report.

[5] Atsuto Yoshizawa, Q&A on Thalidomide-Impaired People, Departments of Emergency Medicine and General Internal Medicine National Center for Global Health and Medicine (NCGM) 1-21-1 Toyama, Shinjuku, 162-8655, Online Publication http://www.ncgm.go.jp/eng/pdf/saridmaid_qa5-5_eng_20140613.pdf

pressure of A. tibialis posterior + 8 mm Hg) ×0.88 mm Hg) for assessment of actual systolic blood pressure by measuring the pressure with a cuff over the arteria tibialis posterior.

I am suggesting the following procedures for blood pressure measurement in thalidomiders with upper extremity damage as a practicable solution:

1.) In cases, where the legs are intact and no risk factors for peripheral arteriosclerotic disease are known and where the individual shows no signs of claudicatio intermittens, intact blood vessels of the legs may be assumed and measurement of the blood vessels in the legs may be used to calculate correct arterial blood pressure.

   a. Upper arm cuff device od "normal diameter" shall be positioned approx.. 3 inch above the ankles in a lying position and pulse of the a. tibialis posterior or dorsalis pedis shall be palpated. Cuff is inflated to the pressure where pulse vanishes. This will yield an approx. 20% higher than correct value and only the systolic pressure can be assessed.
      A results of 120 mm Hg systolic blood pressure in this scenario will mean 100 mm Hg actual systolic blood pressure. Although this will only yield a systolic blood pressure, it may give a rough idea if the blood pressure of the patient is on the rise over the years or not.

   b. Measurement with reference to another patient:
      Blood pressure measurement on lying thalidomide patient on both upper legs and auscultation of Korotkov sounds in the popliteal region.
      Exact documentation of :
      - Position of blood pressure cuff above patella
      - diameter of cuff
      - circumference of knee
      - diastolic and systolic blood pressure results

      - Find healty person, matching in age, height, waist and knee circumference
      - Repeat blood pressure measurement procedure on healthy individual on both legs with identical cuff in same position as in the thalidomide person.
      - take normal blood pressure on both arms of healthy individual as usually.

      Put average arm and average leg blood pressure into relation and multiply the result with documentetd leg pressure of thalidomide person to calculate "arm blood pressure" according to the following formula:

      ASI=LSI*ALI/LLI

      Legend:
      ASI = actual ("arm") blood pressure of short armed individual
      LSI = leg blood pressure of short armed individual
      ALI = Arm blood pressure of long armed individual
      LLI = Leg blood pressure of long armed individual

   c. During surgical intervention, anestesists sometimes use extra large blood pressure measurement cuffs for use on upper legs which permit documentation of systolic and diastolic pressure.

It might be helpful to perform either a simultaneous blood pressure measurement with the anesthesists extra large cuff and the family doctors largest cuff to determine a correction factor or to buy one of these large anesthesists cuffs.

d. Invasive inraarterial measurement of blood pressure during surgery and simultaneous external blood pressure on arms and legs will provide very exact data to use as future correction factor for external blood pressure measurements at the family doctors office. I consider this as the most elegant method and this should be performed when a surgery is done anyway.

2.) In case of arteriosclerotic lesion of the lower blood vessels is suspected, this should be investigated by duplex ultrasound. If suspect turns out to be true, any blood measurement on lower extremities can not be used as base for correct arterial blood pressure calculation without further data (e.G. like described in 1.d.).

As additional investigation I suggest that thalidomiders have their retinas checked with the ophthalmologist once per year to diagnose hypertensive retinopathy as soon as possible. If this is diagnosed in spite of all the blood pressure measurements /calculations having been normal, the blood pressure measurements must be considered as having been falsified. Additional duplex imaging of neck vessels may yield additional information of an undiagnosed hypertensive disease.

Possible new aspects:

24 hour blood pressure monitoring witht the cuff being inflated all 15 minutes has been perceived as disturbing to the extent of being insupportable by the patients and a lot of investigative effort has been invested into finding alternative solutions.
A recent publication (http://www.gesundheitsforschung-bmbf.de/de/4631.php)[6] describes arterial blood pressure being assessed by an ultrasound device and one of the superficial arteries which is compressed from the outside by a small cushion. Beeing a very promising concept in itself, it remains to be elucidated if this can be applied to thalidomiders with an obviously varying anatomical vessel position. A possible solution may be the temporal artery.

[6] 8.12.13: with special thanks to Uwe Zimdars for this information